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2 July 1889.

THE
CEREBRAL PALSIES OF CHILDREN.

THE
CEREBRAL PALSIES
OF
CHILDREN.

A CLINICAL STUDY FROM THE INFIRMARY FOR
NERVOUS DISEASES, PHILADELPHIA.

BY

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DEDICATED

TO

S. WEIR MITCHELL, M.D., LL.D. (HARV.),

PRESIDENT OF THE COLLEGE OF PHYSICIANS, PHILADELPHIA,

IN RECOGNITION

OF

HIS WORK IN SCIENTIFIC MEDICINE

AND IN

GRATEFUL ACKNOWLEDGMENT

OF

INNUMERABLE ACTS OF FRIENDLY SERVICE.

NOTE.—*The material here presented forms the basis of a series of Lectures which were delivered in the spring course at the Infirmary for Nervous Diseases, and which appeared in the 'Philadelphia Medical News,' July 14th to August 11th, 1888.*

CONTENTS.

CHAPTER I.

INFANTILE HEMIPLEGIA.

	PAGE
INTRODUCTION. LITERATURE. HEMIPLEGIA. SYNONYMS. ETIOLOGY.	
Congenital Cases; Conditions of the Mother; Syphilis; Difficult and Abnormal Labour; Injury with the Forceps; Trauma; Ligation of Common Carotid Artery; Infectious Diseases—Scarlet Fever—Measles—Whooping-cough—Cerebro-spinal Meningitis—Dysentery—Vaccinia—Convulsions; Embolic Processes	1—23

CHAPTER II.

INFANTILE HEMIPLEGIA (*continued*).

SYMPTOMS; Onset—Convulsions; Loss of Consciousness; Fever; Delirium; Vomiting. The Paralysis—Face, Arm, and Leg. Rigidity. Reflexes. Sensation. Vaso-motor Disturbances. Electrical Reactions. Post-hemiplegic Movements—Tremor, Chorea, Athetosis. Aphasia—Mental Defects—Epilepsy.	
MORBID ANATOMY. Embolism, Thrombosis, and Hæmorrhage. Atrophy and Sclerosis. Porencephalus.	
DIAGNOSIS. PROGNOSIS	24—55

CHAPTER III.

BILATERAL SPASTIC HEMIPLEGIA.

SYNONYMS. LITERATURE. DEFINITION. SYMPTOMS. CASES. BILATERAL ATHETOSIS—General Description. MORBID ANATOMY	56—75
--	-------

CHAPTER IV.

SPASTIC PARAPLEGIA.

	PAGE
SYNONYMS. LITERATURE. SYMPTOMS. CASES. ETIOLOGY. MORBID	
ANATOMY. DIAGNOSIS	76—87

CHAPTER V.

PATHOLOGY. Hæmorrhage. Apoplexia Neonatorum. Poli-encephalitis.	
Infectious Processes. Peri- and End-arteritis. Thrombosis of	
Cerebral Veins. Condition of the Cord in Spastic Paraplegia.	
TREATMENT	88—103

THE CEREBRAL PALSIES OF CHILDREN.

CHAPTER I.

INTRODUCTION.

DIVIDING the motor path into an upper cortico-spinal segment, extending from the cells of the cortex to the gray matter of the cord, and a lower spino-muscular, extending from the ganglia of the anterior horns to the motorial end-plates, the palsies which I propose to consider have their anatomical seat in the former, and may result from a destructive lesion of the motor centres, or of the pyramidal tract, in hemisphere, internal capsule, crus or pons.

Certain general features define sharply from each other palsies of the upper and lower portion of the motor path. When the latter is affected, as in the common infantile spinal palsy, poliomyelitis anterior, we have the combination of paralysis with rapid wasting, early loss of reflexes, absence of rigidity and marked changes in the electrical reactions. On the other hand, in involvement of the upper segment, when the lesion is cortico-spinal, anywhere from the motor cells of the cerebrum to the gray matter of the cord, there is paralysis with spasm or disordered movements, exaggerated reflexes, neither rapid nor extreme wasting and normal electrical reactions.

The clinical picture presented by diseases of the upper

segment is very varied, depending partly on the nature, partly on the extent of the lesion; and while on certain grounds it would be preferable to classify and consider the affections on an anatomical, or, perhaps, better still, on an etiological basis, we may, for clearness and convenience, adhere to custom and classify the cases according to the distribution of the paralysis, whether hemiplegic, diplegic, or paraplegic. The cases are usually arranged under the generic terms cerebral palsies—the German *Cerebrale Kinderlähmung*—or spastic palsies. while the specific designation indicates the distribution of the paralysis, whether unilateral, bilateral, or paraplegic.

Without entering into historical details, it will be sufficient to note that the publication, in 1884, of Strümpell's paper¹ seems to have aroused special interest in the subject. Since then the monographs of Gaudard² and of Wallenberg,³ the contributions of Ranke,⁴ Bernhardt,⁵ and Kast,⁶ in Germany; of Jules Simon,⁷ Richardière,⁸ Jendrassik and Marie,⁹ in France; of Ross,¹⁰ Hadden,¹¹ Wolfenden,¹² Abercrombie,¹³ and Gowers,¹⁴ in England,

¹ "Ueber die Acute Encephalitis der Kinder," "Jahrbuch für Kinderheilkunde," 1884; and his Text-book of Medicine, by Shattuck, New York, 1887, p. 704.

² "Contribution à l'étude de Hémiplegie cérébrale infantile," Genève, 1884.

³ "Ein Beitrag zur Lehre von den Cerebralen Kinderlähmungen," "Jahrbuch für Kinderheilkunde," 1886.

⁴ "Ueber Cerebrale Kinderlähmung," "Jahrbuch für Kinderheilkunde," 1886.

⁵ "Hemiplegia spastica infantilis," "Virchow's Archiv," Bd. 102.

⁶ "Archiv f. Psychiatrie," Bd. xviii.

⁷ "De la Sclérose cérébrale chez les enfants," "Rev. men. des Maladies de l'enfance," t. i and ii.

⁸ "Etude sur les Scléroses encéphaliques primitives de l'enfance," Havre, 1885.

⁹ "Archiv. de Physiologie," 1885.

¹⁰ "Brain," vol. v.

¹¹ "Brain," vol. vi.

¹² "Practitioner," vol. xxxvi.

¹³ "British Med. Journal," 1887, vol. i.

¹⁴ "Diseases of the Nervous System," London, 1887, vol. ii; "On Birth Palsies," "Lancet," 1888, vol. i.

have extended and systematized our knowledge of these cases.

The valuable papers by McNutt,¹ Sinkler,² Wood,³ Hatfield⁴ Knapp,⁵ Bullard and Bradford,⁶ Seibert, Caillé, J. Lewis Smith,⁷ and quite recently Lovett,⁸ show that the question has not failed to attract the attention of American observers.

It is a pleasure to speak specially of the work of Dr. Sarah J. McNutt, of New York, which Gowers has recently characterized as "by far the most valuable contribution to medical science that the profession has yet received from members of her sex."

The kindness of my colleagues, Dr. Weir Mitchell and Dr. Wharton Sinkler, has enabled me to utilize their cases as well as my own, and makes the material upon which this study is based exceptionally large, much larger in fact than has been heretofore analyzed from any clinic. I have also to thank Dr. Kerlin, of the Pennsylvania Institution for Feeble-minded Children at Elwyn, for placing at my disposal the cases under his care; and to Dr. Wilmarth, his assistant, for anatomical data.

The cases may be arranged as follows :

	Cases.
Hemiplegia { Infirmery	97
{ Penna. Inst. F. M. C.	23
Total	120
Bilateral hemiplegia	20
Paraplegia	11

¹ "Double Infantile Spastic Hemiplegia," 'Amer. Journ. of the Med. Sciences,' 1885, vol. i; "Apoplexia neonatorum," 'Amer. J. of Obstet.,' 1885.

² "Paralyses of Children," 'The Medical News,' 1885, vol. i.

³ "Spastic Infantile Paralysis," 'Polyclinic,' 1886; 'Nervous Diseases; their Diagnosis,' Philad., 1887.

⁴ 'Archives of Pædiatrics,' 1886.

⁵ 'Journal of Nervous and Mental Diseases,' 1887.

⁶ "Report of Proceedings of Suffolk District Medical Society," 'Boston Med. and Surg. Journ.,' 1888, vol. i.

"Report of Proceedings of New York Academy of Medicine," Jan. 25th, 1888; 'Journal of the American Medical Association,' Feb. 25th, 1888.

⁸ 'Boston Medical and Surgical Journal,' June 28th, 1888.

A total of 151 cases of cerebral paralysis.

It is of interest to compare the relative frequency of the cerebral and spinal forms of infantile paralysis. During the period in which there have been at the Infirmary about 120 cases of the former, there have been nearly 500 cases of the latter, so that the proportion is about one to 4.16.

INFANTILE HEMIPLEGIA.

SYNONYMS.—Hemiplegia spastica cerebialis (Heine). Hemiplegia spastica infantilis (Bernhardt). Acute Encephalitis der Kinder (Strümpell). Die Atrophische Cerebrallähmung (Henoch). Agénésie cérébrale (Cazauvieilh). Sclérose cérébrale, atrophie partielle cérébrale (French writers).

ETIOLOGY.—Of the 120 cases, fifty-seven were boys and sixty-three girls. Right hemiplegia occurred sixty-eight, left in fifty-two cases.

Age at onset.	Cases.
Congenital.	15
1st year	45
2nd „	22
3rd „	14
4th „	1
5th „	3
6th „	3
7th „	3
8th „	1
9th „	1
10th „	1
Above 10	1
	<hr/>
	110
Age at onset not given	10

Thus the greatest proportion of cases occur during the first three years of life.

Of the *congenital* cases, five presented no record of injury during delivery, and the affection was noticed

either just after birth, or very early, without definite onset.

CASE 2.—Joseph C—, æt. 2½. H., 203.¹ The mother, when pregnant with him, had chorea from the second to fifth month. When born the left arm was cold and white, and the child never used the left side. A squint has developed during the past six months. The left arm and leg are small, stiff and shrunken.

CASE 35.—Genevieve C—, æt. 2¾. I. P., 4, 38. Eleventh child, birth normal; no convulsions; dentition normal. After birth, the mother noticed that the child used the left hand rather than the right, and this condition has persisted. The child now walks, but with a hemiplegic gait. Rigidity at the right elbow; contractures; difficult to extend. Intellect defective.

CASE 43.—Tillie N—, æt. 23 months. I. P., 3, 185. One of seven children; others healthy. Mother noticed since childbirth a contraction of fingers of right hand, and loss of power in arm; also weakness in right leg, especially at ankle. No history of fever. Born naturally at full term.

CASE 72.—Henry B—, æt. 16. H., 262. Had seven spasms during the first twenty-four hours after birth, followed by loss of power on the right side. Began to walk at twenty-two months. Learned to talk easily. Had a talipes equino-varus of right foot, which was operated upon. Began early to have choreic movements of the right side. Goes to school and is intelligent. The right arm is apparently as well developed as the left, but the muscles are rigid. Very little voluntary motion. At rest, there is frequent spasm of the arm, the fingers are thrown out into irregular movements, and the arm is thrust out at right angles, or jerked behind him. This is very marked on excitement. The right leg is an inch shorter than the left, and not so well developed. When walking the leg is very rigid, and he turns the foot out.

¹ The letters and numbers after the cases refer to the Hospital Case-books.

The leg is frequently extended when at rest. No rigidity of muscles of neck or face.

CASE 73.—Minnie C—, æt. 8. H., 281. Mother noticed that as soon as the child began to crawl and play it seemed paralyzed on the right side. Cannot fix the date of onset; there have not been convulsions. The right arm and leg can be moved, but they are somewhat wasted, and are the seat of irregular choreic movements whenever an attempt is made to use the arm or to walk. There is slight irregular movement on the right side of the face, when talking.

In the first three cases, the condition is stated to have been observed from birth, and, in the last, there was no seizure to indicate the onset, and in it, too, the paralysis was probably congenital. In the fourth case, there were convulsions during the first twenty-four hours similar to those which, as we shall see, usher in the majority of these cases.

Abnormal conditions of the mother during pregnancy, or accidents, are mentioned as possible causes, and, in a few of the cases, we find a record of sudden fright or unusual mental distress; but it is very doubtful how far such influences can be connected with the affection of the child. In such an instance as the following, the fright of the mother early in pregnancy cannot have had the slightest effect in inducing hemiplegia.

CASE 8.—Luther P—, æt. 4. H., 279. Is the fourth of five children. Natural labor. Mother states that she was badly frightened at the third month, and to this she attributes the paralysis of the child. Had spasms while teething, but, as early as the third month, it was noticed that he did not use one side well. The right arm and leg are stiff and contracted. Began to walk when two years old. The speech is somewhat affected. Has had only one convulsion since teething.

Except in a few cases, no special mention is made of the existence of nervous disease in the parents, and the

following is the only case in which there was a marked history of *alcoholism*.

CASE 78.—Florence H—, æt. 7. I. P., 359. Father and mother addicted to drink. Is paralyzed on left side. Can get no history of its origin. Left hand shows marked choreic movements. Gait hemiplegic. Has *petit mal*, and, occasionally, severer fits, in which she falls.

I have been rather struck with the vigorous, healthy-looking condition of the mothers with hemiplegic children whom I have seen at the Infirmary.

Syphilis is not often mentioned as a cause of infantile hemiplegia. It was noticed in only two of Gaudard's¹ series, and Wallenberg² alludes to it, but does not give any instances. In Abercrombie's³ series of fifty cases, four of the children had congenital syphilis. The following is the only case on the records in which there seems to be a pretty definite history of this disease.

CASE 64.—Annie F—, æt. 2. I. P. A., 225. Parents healthy. Of nine children, only two are living. Several died immediately after birth. Mother has cicatrices about the mouth and nose, due to a rash which came on during pregnancy. The child was well when born. When eight months old, fell off a chair; was not insensible, but that evening had "spasms." When ten months old, had spasms while cutting teeth. There were fever and a great increase in the general tenderness which had existed all over the body. There was no coma. About a week after the convulsions the mother noticed that there was complete loss of power in the left arm and leg. The child gradually regained use of the leg, and, when a year old, could stand on the leg, but she cannot yet stand alone. There was no facial palsy. *Status præsens*: A pale, but plump child. Is not very intelligent; can say only two or three words. She has thirteen teeth, all beginning to decay. The left arm is not wasted, but is

¹ Gaudard, loc. cit.

² Wallenberg, loc. cit.

³ Abercrombie, loc. cit.

flexed and contracted, and cannot be moved. The fingers are clenched. The left leg is swollen and softer, but there is no difference in the length. Four months ago she began to have spasmodic contractions of the affected arm and leg, lasting for about five minutes, and occurring several times each day. The arm would be jerked up, the leg twitched and the eyes become fixed. No twitching of face. Just before they come on she gets quiet, and, if nursing, drops the nipple. Immediately after, she draws a deep breath, and is then as bright as ever. The child was brought back when eight years old, and note merely says, "Decidedly idiotic."

The association of paralysis and mental defects with *difficult* or *abnormal labor* has been insisted upon by many writers, and, on several occasions, Dr. Sinkler¹ has called attention to the subject, in connection with cases brought to the Infirmary.

In two cases the children were born prematurely. In Case 51, the mother had a fall, and delivery occurred at the eighth month. The child, however, thrived until the second year, when it had convulsions and became paralyzed on the right side. There was, probably, no connection whatever between the premature delivery and the onset of the disease, but, in the following instance, the affection probably dated from a few weeks after birth.

CASE 66.—Andrew S—, æt. 1. A seven months' child; very thin when born. When three weeks old had many convulsions, very severe. For six months, on and off, there were fits every day. When lifted, and the legs stretched, always screamed. When about five or six months old it was noticed that the left arm and leg were not moved. There has been no improvement, and now the fingers are contracted, and the forearm flexed on the arm. Can raise the arm, but does not use it. Left leg

¹ Sinkler, Discussion on Dr. Parvin's paper on Injuries of the Fœtus, 'The Medical News,' 1887, ii; "Palsies in Young Children," 'Amer. Journ. Med. Sciences,' 1875; 'The Medical News,' 1885, i.

as long as right, but the foot seems shorter. Limb flexed at knee, but he can move the leg and toes. Does not sit alone nor stand. Died a week after first visit.

In three of the cases of bilateral hemiplegia the children were born at the seventh month; and in two of the cases of paraplegia the delivery was premature. In twenty-eight of Little's forty-nine cases of spastic rigidity,¹ eight of which were hemiplegias, the birth was premature, in either the eighth or ninth month.

*Injury with the forceps.*² Fissures and fractures of the cranial bones, with hæmorrhage or contusion of the brain substance, are well recognized by obstetric writers as among the untoward results of forceps delivery. The parietal bones are most frequently involved. Unless bleeding occurs, or contusion of the subjacent cortex, the effect is not serious. In the special monographs relating to cerebral palsies in children, we do not find many cases of the kind. Thus, in Little's³ paper, which deals particularly with the relation of abnormal parturition to physical defects in the child, there are only four instances in which the forceps was used, and there is no statement of actual injury. Gandard,⁴ in a review of eighty cases, met with no observation of the kind in the literature, and Wallenberg,⁵ in his analysis of 160 cases, notes that in only six instances was difficult labor mentioned as a cause, and he says nothing about the forceps. The Infirmary

¹ Little, 'Obstetrical Society's Transactions,' vol. iii, 1862.

² With the revelation of the Chamberlens' secret in the second or third decade of the eighteenth century, and the general introduction of the forceps as an aid to delivery, there was, very naturally, discussion upon the effect of such a measure on the child's head; and to this Sterne gives popular expression when, in 'Tristram Shandy' (vols. i and ii of which appeared in 1759), he makes the breaking of Tristram's nose by Dr. Slop's forceps the beginning of all his troubles. The possibilities of injury to the "delicate and fine-spun web" of the brain are discussed at length by Shandy, sen., with the Doctor and Uncle Toby.

³ Loc. cit.

⁴ Loc. cit.

⁵ Loc. cit.

records contain the following cases, nine in number, in which the children were delivered with forceps.

CASE 6.—Ada W—, æt. 3. H., 249. No other children. The mother had a convulsion during labor and the child was born with instruments. Very early it was noticed that she did not use the right arm and leg properly, and this continues.

CASE 12.—Albert McM—, æt. 18 months. H., 295. Instrumental labor; head slightly injured. Paralysis noticed October, 1884, just after a fall. Whole left side is affected; contractures of arm and wrist. Has no convulsions.

CASE 16.—Mildred M—, æt. 3 months. H., 326. The second child. Was born with instruments. Labor tedious, head locked. Has no marks; well-nourished, healthy child; head symmetrical, a little fuller right than left. Legs never affected, reflexes good. Left arm paralyzed, smaller than right, three-quarters of an inch less in circumference of forearm. Fingers contracted. Improved somewhat under treatment.

CASE 19.—Mary C—, æt. 1. I. P., 49. Parents healthy; was born with forceps. Baby did not use the right arm. Now is unable to sit up. Right arm is smaller than the left, the flexors are contracted, and the fingers are contracted and stiff. Uses the left leg more than the right. There is strabismus; forehead is narrow, no marks; has never had fits.

CASE 47.—Clarence H—, æt. $3\frac{1}{2}$. M., 5, 314. Elder of two children. Other five months old, well. Born with forceps; bears marks of deep lacerations. At two years began to walk, but badly. Walked in a stooping position and on his toes. Momentary spasm six months ago, thought to be brought on by excitement. Fairly well nourished and very intelligent; speaks slowly. Walks with uncertain, tottering gait. Drags left leg, and often falls forward.

CASE 65.—Estella M—, æt. 3. I. P. a., 227. Was delivered with forceps after a long labor; head slightly

cut. No convulsions, but for the first three months of life had prolonged screaming spells. Mother does not know whether the child was paralyzed at birth, but when she was three weeks old, she noticed that the left arm and leg seemed perfectly powerless. When one year old, she began to use the leg. Has now paralysis of left arm; can lift it, but the hand is practically useless, though she can move the fingers. The left leg is very little smaller than the right. Gait hemiplegic. Intelligent.

CASE 67.—Kate F—, æt. 4. M., 6, 18. Born with instruments; bears mark on face; small spot in skin and nodular thickening on right frontal bone. Never has used left hand and arm and left leg properly; cannot grasp well with left hand. Left hand smaller than right; cannot pick up small objects with the fingers; can walk and run, but not well. Left foot stiff; flexion and extension difficult. Apparent rigidity of ankle-joint. Muscles well developed. Walks on outer side of foot, and chiefly on the ball of foot. Knee-jerk increased very little. Left leg is colder. Is bright and intelligent.

CASE 81.—Floyd S. L—, æt. 6. I. P., 33. Supposed to have been injured at birth by forceps. Was very inert during the first three months of life. After this, the mother noticed peculiar spasms, first in left leg then in right arm. Got well except in the right arm, which is choreic and palsied.

CASE 97.—Lulu H—, æt. 5; the fifteenth child. Others born living. Four children had died, two of convulsions. Mother in labor three hours; forceps applied, as there was some difficulty. Child resuscitated with great difficulty. Bears the scars of the forceps on right temporal region, just within line of the hair, and the top of the right ear is scarred. On the left occipital region low down is a scar. The wounds did not bleed much. On the second day child had a convulsion, lasting many hours; never has had another. When about three months old, it was noticed that the left hand was not used, and

that the face was crooked ; and at the eighteenth month, when she began to walk, the left leg was seen to drag. *Status præsens* : Well-nourished, intelligent-looking child. Head well shaped. Slight scars in above-mentioned regions ; bones not apparently injured. Left face atrophied, but the muscles move quite well. Left arm not used ; is smaller than the right. Hand not contracted ; fingers can be flexed. A little stiffness at the elbow. Gait hemiplegic, left leg dragged ; it is a trifle shorter and is smaller than the right. The reflexes are not exaggerated. Knee-jerk not obtainable on either side.

In six of these cases the child is said to have been injured by the forceps, and in all the paralysis was either noticed at once or a few months afterwards without definite onset. Only one of these cases (47) appears to have had spasms.

Trauma.—In three cases there was a history of injury to the head, one a penetrating wound, and two the result of falls.

CASE 23.—J. E. K—, æt. 27. I. P.₁, 131. When eleven months old received a wound on the left side of the head from a pitch-fork, which penetrated the skull from one to two inches, causing immediate paralysis of the right side of the body. Had convulsions after it. In fourteen days began to recover. Was five years before he could walk, and he never has regained the use of the right arm, which is rigid and flexed. Leg is slightly contracted at the knee and is stiff in walking.

CASE 62.—Thomas McK—, æt. 3. I. P.₃, 355. Parents healthy, birth normal. Several children have had chorea. When six or seven months old, child fell downstairs, striking her head constantly. Some time after, the mother noticed that there were irregular movements in the right hand, and the child has never used it properly. The movements are much increased by voluntary efforts.

CASE 79.—Wilhelm S—, æt. 29 months. I. P. a, 81. When born healthy. On the tenth day after birth the mother fell downstairs with him, fourteen steps. The head was not cut or bruised. For eight days he seemed very ill and did not take the breast; did not have convulsions. The head became much swollen—"all out of shape." Has never used the left hand since the fall, and the muscles are now in condition of rigid spasm. Left leg not used so well as right. The right parietal region bulges, the left is flattened, which makes the head very shapeless—right semi-circumference nine and one-half inches; left, eight and seven-eighths inches. There is a large soft rachitic spot on right parietal eminence, very tender on pressure. Fontanelles closed. Has not had any convulsions.

Ligation of common carotid.—In Case 7 of the Elwyn series there is the following remarkable history:

Mary P—, æt. 24. In 1869, when six years old, she had an extensive abscess of the neck following scarlet fever. Ulceration of the right carotid occurred, necessitating ligation, which was performed by Drs. Keys and Getchell. Left hemiplegia followed and has persisted. *Status præsens*: Well-grown but slightly-built woman. Left hemiplegia. Wrist flexed immovably at right angles; thumb held in palm; fingers flexed, but can be moved a little. Arm somewhat wasted. Drags left foot. Left leg a little wasted. Face not affected. Is bright and intelligent. Is not epileptic.

Infectious disease.—It is well known that both cerebral and spinal palsies may follow any of the specific fevers. Gaudard mentions whooping-cough and diphtheria among the possible etiological factors. In Wallenberg's statistics of 160 collected cases, nine are stated to have followed measles; thirteen scarlet fever; three diphtheria (and croup); six epidemic meningitis; three whooping-cough; four typhus (abdominalis); and two vaccinia.

Marié¹ has reported two cases illustrating the connection between infantile cerebral hemiplegia and infectious diseases ; one came on with whooping-cough, the other followed mumps. Abercrombie lays special stress on the importance of this factor, which is noted also by Strümpell, Bernhardt, Gowers and others. In our series, in sixteen cases the disease came on in connection with, or just after, an attack of one of the infectious diseases.

Scarlet fever : seven cases.

CASE 4.—John K—, æt. 12. H., 231. Family history good. Was well until July, 1880, when he had a bad attack of scarlet fever ; ill for two months and had dropsy. Became very thin after the dropsy subsided ; had retention of urine. Convalescence slow—had otitis media and a suppurating cervical bubo. The mother noticed loss of power on the right side as the dropsy disappeared. This persists, and the leg is now weak. But both are getting stronger.

CASE 13.—Emma G—, æt. 17. H., 300. Was healthy as an infant and child. Has not yet menstruated. When fifteen, had scarlet fever, a severe attack followed by dropsy. Was ordered a warm bath and while in it was seized with paralysis of the right side and loss of speech, and was unconscious thirteen hours. Was in the bath three-quarters of an hour. Had fever, which lasted three days. Was in bed for a month. Gradually recovered use of leg. Gait is hemiplegic. Arm stiff at elbow and wrist, and is held flexed. Reflexes exaggerated on right side ; speech is still a little thick ; a little paresis of right corrugator supercillii. Apex-beat of heart forcible ; loud blowing systolic murmur.

CASE 17.—Wm. Mc—, æt. 4. M., 37. The seventh child ; six years elapsed between sixth and seventh. Was well until November, 1884, when he had scarlet fever ; had dropsy and also a very bad throat with it. Shortly after, he had convulsions and became paralysed on the right

¹ 'Progrès Médical,' 1885, No. 36.

side ; recovered power gradually, but has never spoken since. Said a few words just after the spasms. Is a strong well-built child. No note as to spasm of the right side.

CASE 24.—John W—, æt. 4. I. P., 139. Was healthy when born ; other children healthy. When ten months old could walk and was quite well ; was attacked with summer complaint, which left him weak for several months. When two years and three months old, had a *scarlet rash*, from which he recovered and was walking about. Two weeks after, he arose one morning all right, seemed well and took his breakfast. Went to sleep in the morning, as usual, and the mother noticed that he began to pant and the mouth was drawn to the right. For a week he had a series of convulsions and lost power completely in right side and could not speak. In four or five months the speech gradually returned. Began to walk last spring. Convulsions have not returned. Intelligence below par ; talks, and his memory is good. Right arm feeble and contracted, and the fingers are flexed. Gait hemiplegic and the leg is stiff.

CASE 28.—Cassie McA—, æt. 20. I. P. b., 85. Small, thin, pale girl. When between seven and eight years old had a severe and prolonged attack of scarlet fever, during which she was for a time insensible. Does not know if she had fits. Right-sided hemiplegia came on during the attack and has persisted ever since. The face has improved, but the arm and leg have not grown, and there is a marked spastic condition of the muscles. Muscles respond to induced current.

CASE 70.—Alice R—, æt. 20. H., 195. Was imperfect in some way at birth ; the head was bandaged as she was supposed to have water on the brain. She got better ; no special feebleness noticed at that time. At ninth month, scarlet fever ; feebleness on right side followed. Did not learn to walk until three years old ; at first seemed to walk all right, but soon noticed that she turned the right ankle out, for which a brace was applied. At age of ten, the right tendo Achillis was cut. There was

a gradual leaning to the right side, with curvature of the spine. The right arm and leg did not seem to grow proportionately to the left. *Status præsens*: Is emotional. Slight loss of power on right side of face and uncertainty of movements, which are jerky. Great incoördination on attempting to pick up objects with right hand; movements choreic and weak. Some loss of feeling in right hand. Sensation better in right leg than in right arm. Temperature lower on right side than on left. Measurements of right arm and leg, from one-half to one inch less than left.

CASE 83.—Annie K—, æt. 5. I. P.₃, 187. Third child. Others healthy. Family history good. When two and a half years old, in October, the child had a mild attack of scarlet fever, followed by whooping-cough; one day the mother noticed at table that the child had suddenly lost power in the leg and arm of the right side, and that the face was drawn. This was quite sudden, without any premonition. The paralysis persisted, though improving. *Status præsens*: Opens and shuts the right hand, but does not use it habitually. Movements of the arm and forearm good, but constant choreic motion, and it is thrown about, particularly behind her, as she walks. The right foot strongly inverted; gait hemiplegic; toes flexed. Walks on the toes and wears out the nails. No special atrophy of the right side.

Measles: four cases.

CASE 32.—Richard B—, æt. 25. Con. Dis. 2, 215. Was healthy until eighteen months old, when he had measles, followed by otitis, during which fragments of bone came away. He then had convulsions and left hemiplegia, which latter has persisted. The arm is short and stiff—the forearm much shorter than its fellow. Great stiffness; claw-hand. Hemiplegic gait. He has a little difficulty in speech. Intellect is pretty good. The convulsions became worse about the tenth year, and he now has attacks at intervals of a week or two.

CASE 82.—John C—, æt. $2\frac{1}{2}$. I. P.₃, 55. Is the fourth of five children. Cut teeth early. Was well and healthy until last autumn, when he had a spasm lasting four hours. Convulsion general. Next day seemed bright and well. Six weeks ago the other children had measles, and this one, on the second day of the eruption, had a convulsion lasting six hours, after which there was right hemiplegia, with loss of speech, which continued two days. Then convulsions began again, with less severity; sometimes more marked on one side than on the other. He had, in all, about eighty fits. In about two weeks there was improvement in the paralysis, which has continued, and he has, for the past two weeks, been walking. No fits since the eruption disappeared. A discharge from the ear began six weeks ago; has now stopped. Appears to understand what is said, but not fully. Head large and square; slight depression at anterior fontanelle, which is closed. Is irritable. There are distinct choreic movements of right arm, which he uses only when the left is held. There is evidently loss of power. Leg seems to have recovered perfectly.

CASE 37.—Bessie K—, æt. 3 years and 2 months. I. P.₄, 73. Well until the eighteenth month; could walk. During the early stage of measles was seized suddenly with spasms and was unconscious for eleven hours. Had vomiting; paralysis of right side noticed immediately after and loss of speech. Has recovered use of leg and can run. Arm affected. Cannot talk. Intelligence not good.

CASE 1 (Elwyn series).—Jennie S—, æt. 14. In institution five years. Was well and strong as a baby until the twelfth month, when she had measles, followed by paralysis of the right side. Is intelligent. Arm stiff; not much wasted. The fingers move irregularly at times. Under the influence of emotion the hand gets very stiff, but when she is quiet it relaxes and she can pick up objects. Legs well nourished; very rigid; drags much in walking. Knee-jerk not obtainable. No epilepsy.

Whooping-cough: three cases.

CASE 83.—Given under scarlet fever, which preceded the whooping-cough.

CASE 85.—Kate B—, æt. 7. I. P.₃, 248. Has been a healthy child. Last winter had whooping-cough, which lasted into the spring. In May, 1883, had a convulsion, suddenly, which lasted three hours and was followed by paralysis of the right side and aphasia. Did not attempt to walk for two months. Right arm improved, and face did not seem much affected. Gait is hemiplegic. Articulation is now perfect. Can flex and extend the arm and forearm, but has no power over the fingers. Thumb strongly adducted; wrist flexed. There is at times large tremor of the whole arm, especially when at rest. The right leg is stiff, but she drags it. The toes are turned in, but she gets along pretty well. Intelligence unimpaired.

CASE 6 (Elwyn series).—John D—, æt. 14. Difficult, instrumental labor. Well until two years old, when he had whooping-cough, followed by brain fever and left hemiplegia. Arm stiff and wasted; hand flexed; irregular movements in fingers. Leg wasted. Gait hemiplegic. Choreic movements in left facial muscles. Is a low-grade imbecile and has epilepsy.

In the following case *cerebro-spinal meningitis* was stated to be the cause of the trouble, but it was most probably a mistaken diagnosis, as the symptoms are just those which occur in the majority of the cases.

CASE 21.—Lily H—, æt. 4. I. P.₁, 85. Other children healthy. Was normal as a baby. When nineteen months old had an attack which was called cerebro-spinal meningitis; a series of convulsions for eleven days, on recovery from which the left side was paralyzed. When seen again, at the age of six years, there were marked arrest of development of the left arm and spasm and contractions of the paralyzed side.

In one instance the attack came on after *dysentery*.

CASE 22.—Gussie H—, æt. 6. I. P.₁, 125. Was never

a very healthy child. When fourteen months old had a dysentery, after which she had two severe convulsions and coma for several hours, and seemed unconscious for six days. The left side was convulsed for three days. When she roused, it was noticed that the left side was paralyzed—face and extremities. In about three weeks the paralysis began to improve. In three months she began to walk, and within a year could walk alone. No convulsions since on left side, but one general convulsion. Talks with difficulty. Left-sided spasmodic paralysis. Gait hemiplegic.

Vaccinia : There are two instances in the literature in which the hemiplegia came on during vaccination. One is given by Heine in his work on *Infantile Palsy*.¹

In Wuillamier's thesis² the following case is given :

J. L— ; well until ninth month ; vaccinated, and toward the close of the process there was fever, with a general papular eruption. Convulsions came on, followed by loss of power on the left side. Subsequently, epileptic attacks and feeble-mindedness.

The following case I saw with Dr. Morris J. Lewis.

CASE 94.—A. L—, æt. 7 ; a seven months' child, vaccinated by Dr. Gerhard when she was about four months old. During the height of the vaccination there were convulsions, chiefly on left side, which occurred very frequently and were followed by deep coma. It was some time before the paralysis was noticed—some months. Now she has left hemiplegia. Arm wasted ; contractions. Gait hemiplegic. Intelligence defective ; speaks badly, but she is learning rapidly under systematic instruction.

Convulsions : In five or six cases the children had had convulsions repeatedly before the onset of the hemiplegia, and it is possible that they caused the lesion on which the paralysis depended.

¹ 'Spinale Kinderlähmung,' 1860, Zweite Auflage.

² 'De l'épilepsie dans l'Hémiplégie spasmodique infantile,' Paris, 1882.

CASE 26.—Bruce B—, æt. 9. I. P.₁, 207. Has had fits since the age of two years, at least once a month. They were general, and he fell in them. Eighteen months ago, after an unusually severe attack, with seven hours of unconsciousness, he awoke paralyzed on the right side ; speech not affected ; no loss of sensation. In three weeks he began to use leg. Has not had convulsions for a year. There is still difficulty in using the arm, which is stiff, and the movements incoördinate.

CASE 39.—C. M—, æt. 3 years and 3 months. I. P.₃, 237. Mumps two years ago ; has had spasms ever since. One week before Christmas had a sharp spasm, which was followed by right hemiplegia. Unconscious for six hours after attack. Could not walk for two or three days. Has not had spasms since. Leg and arm have improved. Rigidity of arm increased by passive motion. Can extend and flex arm ; can extend but not flex fingers. Walks lame on account of contraction of extensors.

CASE 68.—John F—, æt. 7. I. P. b, 183. Mother died suddedly ; six brothers and sisters, all healthy. Commenced having fits at three years. Has convulsions every six or seven weeks. When six, had a very severe fit, lasting two or three hours. Worked all the time. Fit lasted until 12 p.m. Awakened with a "screech." Lost hearing and speech. For ten days he moved his whole body. Had convulsive movements and contractions. When he began to improve, noticed he could not move right hand. Health excellent. Mind much impaired. Does not move right arm. Nutrition about equal. Right hand colder. Grinds his teeth constantly, night and day. Right arm below elbow smaller than the left. Muscles of right arm contracted, will yield, but immediately return. Plays, amuses himself. Takes no notice of others. Right leg a very little shorter than left. Right foot much colder than left, but muscles firm. Tendency to varus in right foot.

CASE 60.—S. S—, æt. 12. C. D. C., 57. Family history good. Healthy and strong until sixth year ;

intelligence very good. Six years ago mother noticed a twitching of left side of face, lasting not more than one minute, and only once a day. Had had no fall, no sickness of any kind. Soon the attacks of twitching of face became more frequent, two or three daily. Head always drooped to left side, and he seemed unconscious, but immediately afterward returned to play. More frequently attacks of twitching of face at night. Two years after first attack of twitching of face, had a general convulsion of left side; was lying asleep at the time, and attack lasted only a few minutes; did not wake up. Next morning early, while asleep, had another attack, which lasted one hour; got up immediately afterwards, and seemed as well as ever. The attack was confined to left side; was paralyzed completely for one month after this; loss of speech. Paralysis suddenly left, and power to speak returned. The convulsive attacks have continued at the rate of from one to twenty per diem, and always during sleep, never when awake, generally at night, but, if asleep during the day, may have an attack. Face flushed during attack, afterwards pale. Never bites tongue nor foams at mouth. These attacks continued to increase until two years ago. At this time began to lose power in arm; weakness extended to legs and, finally, could not move head, and lost power of speech. No palsy of bladder or rectum. No loss of sensation or hearing. In one year power began to return in hand, and gradually returned to legs and head. Still speaks very indistinctly. *Status præsens*: No paralysis of arms or legs. Speaks very indistinctly; mother only can understand him. Mind greatly weakened; and is irritable. Fits occur every night; generally has one to three nightly, and if bromide is neglected will have a greater number. Always voids urine during a fit. Fits last two minutes, and are confined to left side.

CASE 92.—Della W—, æt. 2. C. D.₂, 950. Convulsions began two hours after birth, and have continued ever since. These are of long duration, and affect the right

side more than the left. Six months ago, after a severe fit, she lost power on the right side, and it persists, but is now partial. There are constant choreic movements in the right hand.

These cases are of interest, as confirming Goodhart's¹ view that the convulsions may cause the hæmorrhage, which leaves permanent damage to the brain.

In the following case the hemiplegia came on after violent *vomiting*, caused by eating pokeroot.

CASE 63.—Wm. R—, æt. 13. I. P., 31. Was well until fifth year; at that time ate some pokeroot, which made him violently sick at stomach: the attack lasted all day. After vomiting fell into a profound stupor and was thought to be dead. When he awoke had complete left hemiplegia, entire loss of power, face as well as limbs. Could not move for three or four months. Recovered slowly, leg first, then arm. *Status præsens*: Muscles well developed. Has every motion of left arm and forearm. Can flex hand, but not well, every effort to flex produces more or less extension. Has also lock-spasm of hand; fingers press tightly in palm, extends them by producing extreme flexion of wrist by other hand, which produces reversed condition, and enables him, with aid of other hand, to unlock the fingers. There is choreic movement of hand resembling athetosis. Sometimes hand will not close. All muscles of left arm and shoulder are in constant motion. Muscles rather over-developed. When asleep, spasm subsides completely. Drags leg, but there are no spasmodic movements.

We should naturally think that *embolic processes* had much to do with the production of hemiplegia in children, but neither in the literature nor in our records is there support for this view. In the great majority of the cases the lesion supervenes before the onset of those affections with which endocarditis is associated. Of Wallenberg's

¹ 'Text-book of Diseases of Children,' London, 1887.

cases, only five had endocarditis and embolism. A case reported by Taylor,¹ illustrates the condition very well: A child, æt. 5, with scarlet fever, had convulsions on the fourteenth day, after which a right-sided paralysis remained. There was also partial gangrene of the hand, and dropsy of extremities. The autopsy showed embolic softening in left hemisphere, infarct in kidney and recent mitral endocarditis. Here the connection is very evident, and it is surprising not to find a larger number of cases of this kind. The only instance of embolism on our records is of a similar nature, and is given among the scarlet fever cases. The girl, æt. 15 at the time, had dropsy after fever, and was ordered a hot bath, in which she was suddenly seized with right hemiplegia and aphasia. That this was embolic is likely from the sudden onset, and from the presence, eighteen months afterwards, of a well-marked apex systolic murmur, the result, no doubt, of an endocarditis at the time of the scarlet fever.

In speaking of the morbid anatomy, I shall refer to the sixteen cases, of the ninety autopsies collected in the literature, in which there was vascular obstruction; seven of them were embolic.

¹ 'British Medical Journal,' 1880, ii.

CHAPTER II.

INFANTILE HEMIPLEGIA (*continued*).

SYMPTOMS.—Complex and varied as are the symptoms of infantile hemiplegia they fall naturally into three divisions, those of the onset, those pertaining to the paralysis, and the sequences; and in this order I shall consider them.

The period of *onset* is usually characterized by convulsions and coma, indicating serious disturbance of the brain functions, but there are exceptions to the rule. Thus, in three cases the disease came on *suddenly*, in apparently healthy children, without spasms or loss of consciousness.

CASE 9.—Nellie M—, æt. 12. H. 280. Paralysis came on when she was one year of age, suddenly, with pains; no spasms; whole left side affected; gait hemiplegic; arm and leg swollen and shorter than fellows. Right calf eleven and a half inches, left eleven; biceps, right eight, left seven and three-quarters; dynamometer, right sixty-five, left thirty-five; left side is cold.

CASE 38.—William M—, æt. 4 years and 6 months. I. P.₄, 84. Good family history; was well until three years ago. While playing he became suddenly powerless on right side; had no convulsions, but was comatose for a time; no fever. The face was included and the loss of power was complete. Is a delicate child. Some mental deterioration. Had convulsions ten days before application and now cannot walk.

In Case 77, the hemiplegia came on while the child was dressing one Sunday morning, after an indisposition of a few days' duration.

A *latent*, gradual onset is more common and was noted in ten cases.

CASE 3.—Joseph D—, æt. $2\frac{1}{4}$. H. 219. When six months old the mother noticed loss of power on the right side. This condition persists; the hand is closed, and there is talipes varus of the foot.

CASE 7.—Daniel McS—, æt. 4. H. 253. Natural labor. When one year old it was noticed that he could not sit up and seemed weaker than natural. Has begun to walk within the past few weeks. Has right hemiplegia with contraction of flexors; walks very badly; does not talk, but understands what is said to him.

CASE 75.—Alphonse N—, æt. 14. H. 299. Is one of eight children, five of whom died in infancy. Walked at two years and then walking gradually became impaired. There was no sudden attack. *Status præsens*: Right arm and leg wasted; the right shoulder-joint and blade project, and the whole side looks smaller. The hand is cold and there is marked mottling of skin of legs and arms; choreic movements of right arm, and at times an irregular, jerky motion of the right leg; reflexes present; the superficial are exaggerated on affected side; heart normal.

CASE 53.—Frank H—, æt. 14 months. I. P.₄, 169. Healthy and strong when born. When five months old, while teething, went into a fit, at first very severe, almost every hour for a week, then once or twice a week. Has them now on Mondays, in the afternoon. Noticed feebleness of left side of body; does not know exactly when first observed. There is loss of power on the left side; with contracture of the left arm and hand, with shortening and contractures in left leg. Cannot walk or even stand alone. Improved, particularly the arm, under treatment.

CASE 52.—Henry I—, æt. 5. I. P. b, 37. Parents healthy; healthy at birth; never has used left hand. At eight months observed that thumb was drawn across the hand, fist closed; this became worse. Began to walk at two years, then noticed that left leg was weak. Had a fit at eight or ten months, another a year later, none

since. Mind not developed. Grip good; contraction in biceps, supinator longus and muscles of hand; hand flexed at right angle, can be easily made straight. Arm much smaller than right.

Convulsions.—In the majority of the cases the disease began with convulsions, partial or general. In fifty-two of the ninety-seven cases on our records, this history was given, which corresponds very closely with the figures of Wallenberg: forty-three of eighty-eight cases in which the history was definite; thirty of the eighty cases collected by Gaudard, and in “more than half” of the eighty cases of Gowers.¹

The following set of cases will illustrate the prominence which this symptom deserves in the clinical history of infantile hemiplegia.

CASE 10.—Reta O’N—, æt. 2 years and 4 months. H. 285. When fifteen months old, in April, during teething, had a convulsion; perfectly well before the attack, which came on suddenly and was confined to the left side and lasted eight hours. In July she had a second attack, the convulsions lasting twenty hours. After the April attack there was complete paralysis of left arm, leg, and part of face; no strabismus. The second attack did not increase the paralysis. She had begun to recover power before the July convulsions. Reflexes increased on affected side.

CASE 20.—Francis H—, æt. 5. I. P.₁, 81. Two older children. Healthy child until eighth month, when teething. He then had convulsions, four in number, very violent, which were followed by left hemiplegia. The fits have continued ever since at intervals of a few weeks. Has never walked; intellect feeble; left arm and hand contracted and stiff, but the stiffness can be overcome; the forearm is shorter; stands on the inside of right foot.

CASE 14.—Ella H—, æt. 4. H. 309. Was well until her

¹ ‘A Manual of the Diseases of the Nervous System,’ London, 1888, vol. ii.

second summer. One night had a convulsion, and in the morning was found to be paralyzed on the left side. Gradually recovered, and can now walk well, though stiffly, and the leg assumes various positions while at rest; attempted movements of the arm will bring on associated movements of the leg and foot. In walking the arm is carried stiffly, extended and pointed backward. Voluntary movements of the hand are slow and performed with difficulty; the fingers do not grasp well; reflexes are a little increased on left side.

CASE 15.—Nellie P—, æt. 5. H. 316. One of three children; eldest child had spasms; parents healthy; was natural when born. Convulsions when one year old, while teething; they were confined to the right side and lasted from 8 A.M. to 5 P.M. After this the side was paralyzed. Has had no spasms since. Uses arm awkwardly with some incoördination. The right hand and arm are smaller than left; walks with an apparatus.

CASE 27.—Lewis P—, æt. 16. I. P., 209. Born healthy. At the age of three months was seized with spasms. There is a doubt whether or not they were confined to the right side, but they were followed by partial loss of power on this side. Has never used arm and leg well since, and they are now shorter and in a state of rigidity. The leg is dragged and the gait is hemiplegic; arm and hand very rigid; intellect somewhat impaired.

CASE 29.—Elizabeth W—, æt. 17. I. P. b, 142. Family healthy; child was healthy when born, and well up to twelfth month. Had a slight fit and lay insensible all night. Was thought to have water on the brain and was leeches. Was ill four weeks. Paralysis then came on, involving left side except the face. Leg rapidly recovered power and she walked in a month. The left arm and leg shorter than the right, and the muscles are stiff. Has power of movement in the arm, and the fingers are flexed, and she does not use them. The left arm is two and one-quarter inches shorter than the right, and the

left leg one and one-quarter inches shorter than the right. No difficulty in speech. Intellect impaired.

CASE 34.—Joseph McC—, æt. 2. I. P.₄, 28. Mother very nervous. Attack came on November 7th, 1882, when the child was about a year old. He had vomiting and purging, convulsions and coma, after which there was sudden paralysis of the left side. Face not involved. The paralysis has not improved much. Reflexes increased on left side.

CASE 36.—Agnes H—, æt. 6. I. P.₄, 55. Healthy as a baby; dentition slow. When sixteen months old had convulsions, fever, and sudden paralysis of right side of the body, involving the face. Gradual return of power in face; gait hemiplegic.

CASE 44.—Joseph H—, æt. 18 months. I. P.₃, 189. Natural labor. Child healthy when born. In October was taken suddenly with high fever; no convulsions. The fever subsided in a week. He continued very irritable and nervous for a month. Week before Christmas had convulsions, lasting for fourteen hours, and he was paralyzed in the left arm and leg. The following day unilateral spasm began and continued until evening. He has never had any more spasms. The palsy continues, with, of late, improvement in the leg. *Status præsens*: Weakness and partial palsy of whole of left side with considerable rigidity of flexors of fingers; no deviation of face now, though there was some at first. Died of the unilateral convulsions just eleven months after onset of the disease.

CASE 46.—Mary M—, æt. 19 months. I. P.₃, 165. One other child has had spasms. In July had "spasms." Woke from sleep in them; the left side was convulsed and, after spasm, found to be palsied. Two days later had eighteen attacks; after this left arm and leg seemed a little better. Face was drawn to the left side. After a few days ceased having these violent spasms. Began to walk in two months. Since then she has had momentary attacks of unconsciousness, occurring at first daily,

now every two or three days. *Status præsens*: Healthy looking; no atrophy of muscles; no contractions. Left arm and leg weaker than right. Falls easily. Face not drawn.

CASE 49.—Mary M—, æt. 14 months. I. P.₃, 337. Youngest of three children. Well until July 6th, when she had spasms all the afternoon and evening. No fever or diarrhœa. Said to have had congestion of lungs. Face, arm, and leg right side palsied. Began to move right leg at end of week. Arm still palsied.

CASE 50.—Jennie A—, æt. 14 years and 10 months. H.₁, 348. Was healthy when born. Labor natural. Enjoyed good health until about a year old, when she had convulsions, suddenly followed by coma, which lasted several days, and, on recovery, she was noticed to have lost power on the left side of face, arm, and leg. For several months the palsy persisted, but she gradually regained use of the leg and then of the arm, but the face has never changed; for this she now comes to the hospital. *Status præsens*: A fairly well-grown girl, rather nervous and timid, and the father says she worries herself and has fits of despondency. There is complete paralysis of the lower facial muscles on left side; mouth is drawn to the right. Can shut the eye, wrinkle and elevate the brows, but there is no movement of the lips on left side, nor does the ala nasi expand. Face on this side a little smaller. No loss of sensation. Arm is used quite well; no weakness of hand. Dynamometer: R. 75; L. 65. Can run well; no difference in the legs. Reflexes normal. On watching the face it is noticed that, from time to time, there are choreiform twitchings in the muscles of the face on the left side; most marked when the muscles of the normal side are in action. Quantitative, but no serial changes in electrical reaction of the paralyzed muscles.

CASE 54.—Thomas McR—, æt. 10. Well until eight months old, when he had spasm of the left hand, and the whole left side seemed weak. No statement as to general convulsion. When two years old had seven con-

vulsions, and has had eight since then. Fever often precedes the attacks. There is contracture of the left arm, and the hand is weak, though the fingers can be moved. There are, at times, slight spasmodic movements in pronation. Left leg drawn up by contraction of gastrocnemius.

CASE 55.—Joseph D—, æt. 11. I. P. a, 107. Healthy when born. When three months old had convulsions, after which it was noticed that he did not use the right hand. Had twitchings and spasms as an infant, and, at three years, a severe convulsion lasting from 10 A.M. to 10 P.M. Has not had violent spasms since. Walked when two years old. *Status præsens*: Walks badly on account of contraction of right gastrocnemius. Movements of right arm limited. Cannot extend the wrist; fingers flexed. Grip fairly good.

CASE 57.—Harry S—, æt. 3. I. P. a, 159. Well until the tenth month when, during an attack of catarrh, he had a severe convulsion lasting seven hours, chiefly on right side, and for three days lay in a stupid, semi-comatose condition. Could not use the right arm after it, and the right leg was weakened. The mouth was drawn to the left side, but became straight again. When a year old had a second convulsion while teething. Gait hemiplegic; cannot lift the toes; right leg well nourished. Contraction and spastic condition of the right arm.

CASE 59.—Mary H—, æt. 6. C. D. C., 80. Parents healthy; one brother had convulsions while cutting teeth. Healthy until third year. Fit came on while in perfect health. Fit began on left side, then became general; lasted nine hours. Insensible for some hours. Could not see for two days. Left side completely paralyzed. Could not move left side for three months; improvement beginning in hand. At end of year could walk. In September had fits again, but was not more paralyzed. Fits again in March. Three attacks of fits since. Last attack, one year ago, completely palsied the left side again. Ever since first attack mind affected; bad temper; bites.

No atrophy of left arm, no contracture; left arm seems weak. He walks and runs. Has fits now.

CASE 30.—Christian A—, æt. 8. I. P. a, 19. Healthy when born. When six months old had very violent fits. When three years of age had one day convulsions from 4 A.M. to 7 P.M. Had summer complaint badly during the first eighteen months of life. After the first fit lost power on the right side of the body and in the limbs. Chorea gradually developed. *Status præsens*: Is well grown and intelligent; goes to school. The right arm and hand are weak, and he can flex and extend the fingers, but when he attempts to perform any volitional act, irregular movements come on and prevent their successful performance. The grasp of the right hand is decidedly weaker than that of the left. When at rest the right hand is closed and the thumb flexed across the palm. The right leg is colder than the left and the heel is drawn up by contraction of the gastrocnemius. The foot is inverted, pes equino-varus. There are no choreic movements in the leg. The tendo Achillis was cut and an apparatus applied which relieved him very much and enabled him to walk much better.

CASE 69.—Henry L—, æt. 14. When eighteen months old had general convulsions which lasted ten days, and left the right side paralyzed. Gradually recovered the use of the leg, but the right arm is atrophied and weak, and displays constant choreic movements. No contractions. Walks quite well; general health good.

CASE 88.—Amanda C—, æt. 13 months. I. P.₃, 375. Born without instruments. In October, when seven months old, had spasms and was unconscious from Saturday to Wednesday. Spasms began in left arm and right leg. On Wednesday the arm and leg were noticed to be palsied. *Status præsens*: Is unable to sit alone; falls towards the left side. Rigidity and contractures of left arm and leg, marked choreic movements. Muscles not wasted. The thumb is turned into hand.

Loss of consciousness almost invariably accompanies the convulsions, and may last from a few hours to many days. In a few instances coma occurred without convulsions.

CASE 5.—Lizzie E—, æt. 21 months. H. 237. One of two children. Was healthy when born and until sixteen weeks ago, when, in the evening at 10 o'clock, had a most intense headache. She had seemed to have frontal headache for about two months. She became unconscious and did not revive until 11 o'clock. She attempted to rise but fell to one side. She was able to walk upstairs, and nothing wrong was noticed in the limbs. The next morning the child could not move the right arm or leg, and had lost completely the power of speech. Three days after could move the leg, and walked again in three weeks; regained speech on third day. Has for the past seven weeks begun to use the arm. Patellar reflex is increased on the affected side.

CASE 48.—Bertram T. B—, æt. 5. I P.₃, 279. Family history good. Elder of two children. Two years ago, in July, had an attack of left hemiplegia. Unconscious for six weeks; no convulsions. At end of six weeks began to move right arm and leg. Two months later began to move left leg. Has not improved much since. Considerable wasting on left side, and the left foot is somewhat smaller than the right.

Fever.—The records are not complete on this point, and it is not often mentioned whether or not the temperature was elevated. Fever may be transient or persist for weeks as the following cases illustrate:

CASE 45.—Lewis S—, æt. 2. I. P.₃, 11. Youngest of six children; others healthy. Parents healthy. Well until September. While playing on floor, fell over, and on examination it was found that he did not use left arm and leg well. Paralysis of leg soon became complete and of arm incomplete. Began to use arm in one week, and, in a month, could sit alone and draw up his leg. On

day of attack had fever, was restless, did not vomit. No retraction of head and no bladder trouble. Fever disappeared in a day or two; soreness to touch remained for two weeks. *Status præsens*: Has improved very little in leg. Is a well-nourished looking child; left leg atrophied. No difference in length of legs, but left foot one-quarter inch shorter. Can flex and extend leg and thigh; extends foot, but cannot flex it. Flexes and extends toes. *Talipes equinus*; sensation normal.

CASE 41.—Jennie E—, æt. 4. I. P.₃, 195. In April, had fever and delirium lasting almost four weeks. At about end of fourth week the mother noticed right arm and leg were paralyzed. Speech was also a little affected. *Status præsens*: Right leg a little shorter, about one-quarter of an inch; and foot also. Slight atrophy of leg. Has all movements of leg and arm. Arm slightly atrophied; no change of temperature. In walking she limps, and there is slight “*genu recurvatum*.” There are slight involuntary movements of affected limbs, noticed when excited. September 24th, 1879.—Movements have ceased; walks better, some rigidity of right arm.

Strümpell, Gaudard, and others mention fever as an invariable accompaniment of the convulsions which usher in the disease in such a large proportion of the cases.

Delirium, which is noted by some writers as preceding or replacing the convulsions, was observed in Cases 41 and 84.

Vomiting is especially mentioned in only four cases.

♥ Soreness of the general surface was observed in Cases 45 and 64.

♥ Screaming spells preceded the attack in one case.

Doubtless many children die in the convulsions or in the subsequent coma, without reaching the second or paralytic stage of the disease.

The hemiplegia, which is noticed as soon as the child recovers consciousness, is usually complete. Less frequently there is, at first, paresis which gradually results

in complete loss of power, and in some instances the total paralysis was only established after repeated convulsions, as in Cases 59 and 85. In sixty-eight cases the right side was involved, in fifty-two cases the left.

The *face* is not always affected ; the exact proportion is uncertain as the point was not constantly mentioned. As is the rule in facial paralysis of cerebral origin, the superior muscles are not involved, and the child can close the eyes and elevate the brows. Rapid and complete disappearance of the facial palsy is the rule, and of the cases which returned for examination, in only one did the condition persist, and in this, strange to say, no trace remained of the paralysis of arm and leg. The child, aged fourteen and one-half years at time of observation, had been completely hemiplegic at the age of one year ; the arm and legs had gradually improved, but the face was drawn to the right and there were choreiform movements in the paralyzed muscles of the left side. The tongue did not seem to be affected, but the uvula deviated strongly to the right. Dr. Sinkler has at present in the wards a girl aged thirteen, who became hemiplegic in infancy, and has still complete facial palsy ; and in Case 96, the last of the forceps cases, there were paresis and atrophy of the facial muscles.

As with the hemiplegia of adults, the residual paralysis is most marked in the *arm*, which displays more or less wasting, and is, as a rule, useless for the ordinary purposes of life. There may be arrest of development, leaving a wasted and withered member ; more commonly, there is only a moderate degree of atrophy. In extreme cases the arm is held close to the side, the forearm strongly flexed at right angles and in the semi-prone position ; the hand flexed and the fingers contracted in the palm, usually embracing the thumb. Motion may be almost lost in the arm and completely lost in the fingers. In a majority of the cases there is considerable power of movement, and the arm can be lifted above the head and flexion and extension made at elbow and wrist. The

finer and more delicate movements of the hand are rarely recovered, and while many of the cases possess a useful limb for carrying or grasping purposes, actions requiring manipulation with the fingers cannot be effected. The grasping power is often remarkably good.

The *leg*, as a rule, recovers more rapidly and more completely than the arm, and the palsy may completely disappear, an event which very rarely happens in the upper extremity. The wasting is never so marked, nor does arrest of development often occur. In only one case, 6 of the Elwyn series, is the leg much atrophied. But even when of fair size, and of good muscular development, there is evidence of impaired power in the persistent halt which is such a characteristic legacy of hemiplegia. The patient may simply "favor" the affected side, noticeable, perhaps, only on rapid walking. A very perceptible dragging of the limb is more common. There may be tremor of the leg while in motion.

Rigidity was present in a large proportion of the cases at the time they came under observation. So marked is this feature of spasm that the disease has been termed spastic infantile hemiplegia. Although a very frequent, it is not a constant symptom, and there are cases in which the paralyzed limbs are quite relaxed, even years after the onset of the affection. In Cases 4, 5, and 7 of the Elwyn series the arm is flaccid; and in Case 10 the spasm is very slight. The rigidity disappears during sleep; it is increased by emotion and aggravated by any attempts forcibly to overcome the spasm. When the rigidity is extreme and has lasted for years, permanent contracture may result, in which condition relaxation is no longer possible, on account of the structural changes which take place in the muscle.

The *reflexes* are almost invariably increased in the affected limbs. The knee-jerk is exaggerated, and, in many cases, the ankle-clonus can be obtained. As often happens in hemiplegia, the reflexes may be heightened on the sound side. A rectus clonus can sometimes be got, and, in

several of the cases, making sudden tension of the spastic fingers developed a clonus in the flexors. In three cases the reflexes were absent—Cases 1 and 8 of the Elwyn series, and 9 of the Infirmary patients. In the Elwyn cases there is rigidity, which in the other is very slight. They are not necessarily absent in the instances of flaccid paralysis.

Sensation is not often disturbed; it has been normal in all the cases which I have examined, and it is not once mentioned in the records as absent.

Vaso-motor disturbances, such as coldness of the affected extremities, blueness and congestion, are not uncommon, particularly when the paralysis is extreme.

The *electrical reactions* are usually normal; occasionally quantitative, but never qualitative changes are noted. None of the recent cases examined in the electrical department, by Dr. Willets, have shown alterations other than slight diminution in the response.

Post-hemiplegic movements.—It was from this Infirmary, and based chiefly upon a study of cases of infantile hemiplegia, that Dr. Weir Mitchell first described those disorders of movement which are known as post-hemiplegic chorea, mobile spasm, athetosis, hemi-ataxia and post-hemiplegic tremor. There were thirty-one cases in the entire series which presented these movements. Three varieties were noted:

1. *Post-hemiplegic tremor*, which was present in only one case (85), and occurred in the entire arm, chiefly when at rest.

2. *Post-hemiplegic chorea*, or, more correctly, hemi-ataxia. The great majority of the cases, twenty-four in number, came in this division. The movements were incoördinate and choreiform, chiefly noted on voluntary effort. Many instances of this have been described in the report of the cases.

3. *Mobile spasm and athetosis*, noted in six cases.

The following are good illustrative cases:

CASE 91.—Mary H—, æt. 11. I. P., 161. When two years old, had “a spell of sickness” which lasted some time, and in which paralysis of the left side gradually came on. She did not walk until the fourth year. She now favors the left side in walking, and the arm and leg are stiff. Has slight power over arm, but it and the hand are in constant, irregular motion; fingers are extended in regular order, and she has no control over them. She can move the arm about quite well.

CASE 76.—Edith W—, æt. 6. H., 301. Birth natural, at full term. Father died of phthisis. At four or five months noticed that the child did not use the right arm. Began at two years to pull herself up by a chair, and at two and a half years learned to walk, and then dragged the foot. Has been improving. Face not affected. Is intelligent. Choreic movements of right hand and arm. January 25th, 1888, condition as follows: Is a well-grown, fat child. Walks with a decidedly hemiplegic gait. Sits with hands quiet in her lap, the right on the left. When excited, or on attempting to make any movement, there are regular choreic movements in the right hand and arm. In grasping an object picks it up with the hand, not with the fingers; as the object is approached the fingers jerk in various directions, are separated from each other and extended in more or less orderly sequence. Some stiffness in the arm during these movements. Very similar motions are present in the foot and toes, and the leg is a little stiff. Knee-jerk on right leg exaggerated; no ankle-clonus. Intelligence good.

In one case there were associated movements of the paralyzed arm on attempting motion with the other arm. Allied to this, doubtless, is the not infrequent irregular jerking of the arm in the act of walking. Sometimes it is thrown in a curious way behind the back.

Aphasia.—Speech was affected in thirteen cases, in five of which the children were under three years of age.

The following are illustrative cases:

CASE 1.—Cecilia P—, æt. 12. H., 25. Healthy when born; one of seven; others healthy. Father has had hemiplegia for three months. When two years old awoke paralyzed on right side; became comatose and remained unconscious for two days. Lost power of speech for six weeks. Paralysis persisted for six months; the leg gradually improved. Rigidity came on in affected side a few months after the attack. Has had, at times during the past three years, loss of consciousness, with convulsive movements chiefly of right side. Has had two or three in a day but may not have any for a week or more. Has a characteristic hemiplegic gait. Leg is a little stiff; muscles somewhat wasted; one arm smaller than the other and flexed; the fingers also flexed and contracted.

CASE 11.—Robert M—, æt. 13. H., 91. No injury at birth. One year ago sudden paralysis of the right side, which was preceded for a day by loss of power of speech. The face was also drawn. Had convulsions a few weeks after the hemiplegia, but not at the time. Began to walk eight weeks after the attack, and recovered speech about the same time. Arm has improved but little; is stiff. Convulsions at intervals. Is improving.

CASE 33.—Mathew C—, æt. 10. I. P.₄, 19. Good family. Well until August, 1883, when present trouble began. Had vomiting, diarrhœa, great headache and photophobia, which lasted four days, and on the fourth day there was complete right hemiplegia, including face, with aphasia. In a month the child began to get about; the aphasia was complete for one week, and then the power to walk gradually returned. Walks well but slowly and stiffly. Arm contracted. Right face is still smooth, and the dimple is gone.

CASE 58.—Ellen McC—, æt. 6. C. D. C., 177. Parents healthy. Healthy until three years of age. Then had a fit, lasting, with intervals, for nine days, and, at the end of that time, convulsive movements confined to right side; right hemiplegia followed, and also loss of speech. Gradually regained power of walking; has not yet re-

gained power in arm. Began to speak in a year, and mental condition improved. Ever since first attack has had attacks of unconsciousness every day; jumps, falls to ground and immediately rises again, looking bewildered. No prolonged convulsion until ten days ago. During this time has had none of the lesser attacks. Has had as many as fifty to sixty of these "jumps." Injures herself much by the falls. During attacks, convulsive movements of left side of body and face, and she screams. Does not bite tongue; speech affected since second attack. *Status præsens*: Right arm smaller than left; contractions of flexors of forearm and wrist, not of her fingers. Right leg trifle shorter than left, and slight contraction of tendo Achillis.

Of these thirteen cases twelve were associated with right and one with left hemiplegia. The number affected is much smaller than in the collected series of Wallenberg,¹ whose statistics give forty-five cases of speech disturbance in ninety-four right hemiplegias and seventeen cases in sixty-six left hemiplegias. In Gaudard's² fifty-five right hemiplegias there were twenty-five cases of disturbance of speech, seventeen of which were true aphasia. There were eight left hemiplegias with aphasia. Bernhardt,³ who has made a very careful study of this condition in children, says that in a majority of the cases of cerebral palsy, no matter on what side, there is affection of speech, usually transitory. In one case of our series the aphasia was associated with left hemiplegia. Usually the power of speech begins to return in a short time, but recovery may be deferred for a year (Case 58) and in Case 17 the child had not spoken six months after the lesion. In several instances recovery was incomplete.

In eight of the twenty-two cases at Elwyn, there is absence or profound disturbance of speech, but the patients are idiots or idio-imbeciles who have never acquired fully the power of speech.

¹ Loc. cit.

² Loc. cit.

³ 'Virchow's Archiv,' Bd. 162

Mental defects.—A lesion so serious and extensive as that which is associated with infantile hemiplegia may seriously interfere with cerebral development, and, among the most common sequences, we find various anomalies of intelligence. In the Infirmary series only twelve cases presented idiocy or imbecility at the time of observation, but it must be remembered that the majority of the cases came to the Infirmary a short time after the onset of the paralysis, so that our records on this special point are by no means complete. In Gaudard's series—eighty cases—there were fifteen feeble-minded and nineteen idiotic children, while in Wallenberg's cases (160), there were fifty with mental defects, and in fifteen cases imbecility followed the epilepsy, so often associated with this condition. Three grades may be distinguished: idiocy, which is most common when the hemiplegia has existed from birth, or has come on at an early period; imbecility, which may increase with the development and persistence of convulsions; and a feeble-minded backward condition, a retarded rather than arrested mental development. The twenty-three patients at the Pennsylvania Institution for Feeble-minded Children may be classified as follows: Idiots, four; idio-imbeciles, two; low-grade imbeciles, five; middle-grade imbeciles, six; high-grade imbeciles, six.

In connection with the mental disturbance which so often follows infantile hemiplegia, Merklin¹ has called attention to the development, later in life, of psychoses, even in cases which, in youth, have not shown any abnormalities.

Epilepsy.—One of the most common and distressing symptoms is the occurrence of convulsive seizures, usually confined to the paralyzed side, but tending to become general. This hemiplegic or post-hemiplegic epilepsy affects, sooner or later, a considerable proportion of the cases. In the Infirmary series of ninety-seven

¹ 'St. Petersburger med. Wochenschrift,' 1887.

cases there were only twenty instances. In Gaudard's list (80) there were eleven cases of hemi-epilepsy, and sixty-six in Wallenberg's series of 160 cases. At Elwyn of twenty-three cases, fifteen had epilepsy.

The attacks begin a variable time after the onset of the hemiplegia, mostly within two or three years, though they may be deferred eight or ten years, or even longer. In some instances the paralyzed limbs are convulsed within a few weeks.

These seizures may be either (1) attacks in which the child is simply dazed for a moment or two, occasionally longer without any motor involvement; (2) spasms beginning in and confined to the affected side, without loss of consciousness—the true Jacksonian epilepsy; (3) general convulsions which begin in the paralyzed limbs, and are usually accompanied by loss of consciousness. All three forms may occur in one case, but by far the most common and characteristic is the hemi-epilepsy with retention of consciousness.

The following illustrative cases will give an idea of these attacks:

CASE 96.—Patient of Dr. Mitchell, at present in the house. The hemiplegia is left-sided, and dates from shortly after birth. As the child sits on the floor the eyes will suddenly become fixed, deviate strongly to the left, the head begins to nod (the so-called nodding epilepsy, *E. nutans*). The left arm, which is constantly stiff, may be more closely retracted, but often the fixing of the eyes and nodding motion of the head alone indicate that a fit is present. After a few moments the child wakes up with a start and resumes its play.

In Case 64 the child, *æt.* 2, became hemiplegic (left) at the tenth month. When twenty months old she began to have spasmodic contractions of the left arm and leg, lasting for about five minutes, and occurring several times a day. The arm would be jerked up, the leg twitched, and the eyes become fixed. Just before they come on, she gets quiet, and, if nursing, drops the nipple. With

cessation of the attack she draws a deep breath and is then as bright as ever.

In the following case the lad had post-hemiplegic movements, as well as distinct convulsions, which sometimes became general :

CASE 74.—Charles W—, æt. 13. H., 323. Was healthy as a baby. Birth natural. In August, 1881, had a "sick spell" from Wednesday to Friday. While dressing on Sunday, lost power entirely in left arm, leg, and face. Was in bed two months. He slowly regained power of leg. Since then has had fits. *Status præsens* : Is fairly intelligent; no scar on head. Characteristic hemiplegic gait; drags the left leg; left arm smaller than the right; it is wasted, and possesses very slight power of movement—chiefly flexion. Spastic rigidity of flexors of the wrist, so that the carpus cannot be extended. Often in the day, as many as ten or fifteen times, the arm will stretch out from the side, and the fingers will extend, first the little and then the others in order. Face muscles will also work. He has other attacks, in which the movements may begin as these, and be confined to the arm or extend to the entire side of the body. He never loses consciousness, and never sleeps after the attacks. Has as many as three or four in a day. He has headache, but not worse on one side. Knee-jerk exaggerated on left side.

CASE 18.—Annie E—, æt. 10 years and 6 months, I. P., 157. Three other children. Was quite healthy until the sixth year, and was a very bright, intelligent child. Had a convulsion one night after exposure to intensely cold weather. The spasms were on the right side. They recurred at intervals for a year, always on the right side, and she gradually began to lose power on this side. When about nine years old the convulsions became general, but they began on the right side. They have become more numerous, and she has had as many as fifteen in a day, and even four or five in an hour. *Status præsens* : Dull, stupid-looking girl; marked spastic

hemiplegia of the right side, arm and leg rigid. Fingers strongly flexed in the hand. Heart normal.

MORBID ANATOMY.—Although the clinical features of infantile hemiplegia are as well characterized as those of infantile spinal palsy, our knowledge of the pathological conditions on which the former depends is, in comparison, still very defective. As in hemiplegia of adults, the lesions are variable, and the symptoms may be produced by any destructive process in the cortico-spinal section of the motor tract. It is surprising how few are the observations made shortly after the onset of the paralysis. The great majority of the post-mortems have been made after the hemiplegia has lasted for months or years, when all trace of the primitive lesion has disappeared.

I have analyzed the records of ninety autopsies in cases of hemiplegia coming on in infancy or childhood. The majority of these are contained in the articles of Cotàrd,¹ McNutt, Gaudard,² Richardière³ and Wuillamier,⁴ but I have, as far as possible, referred to the original cases, and have been able to extend the list of the forty-eight autopsies analyzed in 1886 by Wallenberg. The right hemisphere was affected in thirty-nine cases, and the left in fifty-one. It is interesting to note in seventy cases, as bearing upon the prognosis, the ages at which death occurs. Under two years, 4; between second and fifth year, 10; from fifth to tenth, 8; from tenth to twentieth, 18; from twentieth to thirtieth, 12; thirtieth to fortieth, 4; fortieth to fiftieth, 8; above fifty, 6. I have excluded all cases of hemiplegia coming on after the fifteenth year, and, in the great majority, the onset dated from convulsions in the early years of life.

The lesions may be conveniently grouped under three headings:

1. *Embolism, thrombosis, and hæmorrhage.*—In this group, comprising sixteen cases, the patients did not long

¹ Cotàrd, 'Sur l'atrophie partielle du cerveau,' Paris, 1868.

² Loc. cit.

³ Loc. cit.

⁴ Loc. cit.

survive the attack, and a study of the conditions found post mortem might be supposed to throw considerable light on the nature of the initial lesion. In the cases of Gibb,¹ Vernois, and of Valleix,² the condition was congenital. The mother, in Gibb's case, had received a blow on the abdomen, and the child was stillborn, with rigidity of the limbs on the left side. There was a clot in the right hemisphere above the ventricle. In the cases of Vernois and Valleix the labor was difficult, and the paralysis existed from birth. The children lived for forty-nine days and three and a half months respectively, and, in both, extravasation existed in the neighbourhood of the ganglia. In the cases of Wrany-Neurentter,³ Taylor,⁴ Callender,⁵ Kelly Johnson,⁶ Barlow,⁷ there was heart disease, with embolism of the middle cerebral artery. A case of Henoch's⁸ was also embolic. In two cases of Callender,⁹ in one of Lewkowitsch, and in one of Reimer, there was hæmorrhage. In a case of Abercrombie,¹⁰ the right Sylvian artery was plugged by a firm thrombus. In Case 12 of a series of aneurisms of the larger cerebral arteries, which I reported from the Montreal General Hospital¹¹ a boy of six years, with left hemiplegia, there was hæmorrhage into the longitudinal fissure, and laceration of the median surface of the right hemisphere, caused by rupture of an aneurism of the right anterior cerebral artery. In a case of Dulles,¹² in a child of six months, there was hæmorrhage into the ventricles.

¹ Gibb, 'Lancet,' 1858.

² Quoted by J. Lewis Smith, 'Diseases of Children,' 4th edition.

³ Quoted by Wallenberg, loc. cit.

⁴ 'British Medical Journal,' 1880, ii.

⁵ 'St. Bartholomew's Hospital Reports,' vol. v.

⁶ 'Medical Times and Gazette,' 1880. (Gaudard, Reference incorrect.)

⁷ 'British Medical Journal,' 1876.

⁸ 'Vorlesungen über Kinderkrankheiten,' Berlin, 1883.

⁹ Loc. cit.

¹⁰ 'British Medical Journal,' 1887, i.

¹¹ 'Canada Med. and Surg. Journal,' 1886; 'Trans. Path. Soc. Philadelphia,' vol. xiii.

¹² 'Philad. Med. Times,' 1876.

Thus, of the sixteen cases, there was plugging of a Sylvian artery, usually embolic, in seven, and hæmorrhages in nine. In striking contrast to the majority of the cases of infantile hemiplegia, is the significant fact and in this group the age at onset is high. Excluding the three congenital cases, there was only one child under three years, while ten were over six.

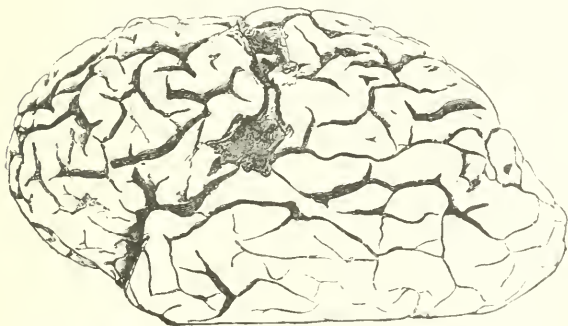
2. *Atrophy and sclerosis.*—In fifty cases there was wasting with induration, either of groups of convolutions, an entire lobe, or, in some cases, the whole hemisphere was affected.

The following cases from the institution at Elwyn illustrate this condition in a typical manner :

Male, æt. 16. Family history good. During measles when a child, he became paralyzed on the right side, and had aphasia. He regained partially the use of the leg, but did not recover speech. There was complete loss of power in the arm. He had repeated convulsions, in one of which he died. There was no sensory disturbance.

The only lesion found was in the left hemisphere. The brain was of full size, and looked natural ; the membranes stripped off readily, except in the Rolandic region on the left side, where the pia mater was greatly thickened and

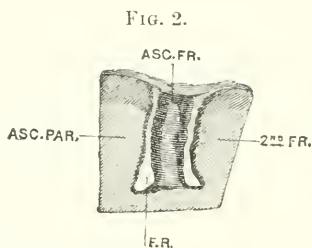
FIG. 1.



Left hemisphere, showing sclerosis in the Rolandic region.

adherent. As shown in the cut, Fig. 1, which is taken from a photograph by Dr. Wilmarth, there is a depression in the neighbourhood of the fissure of Rolando, and the fissure itself is not evident except at its upper third. So far as can be made out, the ascending frontal convolution in the greater part of its extent is atrophic, reaching to its lower end, but above there is at least half an inch unaffected. The central portion of the ascending parietal is also involved. From these regions the thickened membranes could not be removed without tearing the brain substance. The paracentral lobule, the third frontal, and the insula did not appear to be affected. The cord, unfortunately, was not examined.

Sections across the sclerosed area, Fig. 2, show a complete atrophy of the ascending frontal gyrus and of the



Section through the sclerosed area. *Asc. Fr.* Ascending frontal gyrus.

Asc. Par. Ascending parietal. *2nd Fr.* Base of second frontal.

F. R. Fissure of Rolando.

contiguous surfaces of the adjacent convolutions. The pia is thickened and closely adherent, particularly where it dips down to the top of the wasted convolution. In the fissure of Rolando, a loosely arranged connective tissue, containing numerous blood-vessels, unites the opposed pial membranes. At the bottom of the fissure there is a wide sulcus, containing several large veins. In the shrunken ascending frontal convolution three layers of tissue can be readily distinguished, even with a low-power lens: (1) Beneath the pia is a narrow layer, re-

sembling closely the normal first layer of the cortex, a granular matrix with a few nuclei arranged centrally, and, here and there, spaces, probably vascular. Blood-vessels pass through this part from the meninges. (2) A wider, more loosely arranged layer, with numerous nuclei and distinct fibres, among which are small arteries with thickened walls and dilated capillaries. In the deeper portion of the gyrus there are irregular spaces, some of which are large enough to be seen with the naked eye. (3) A central stem, composed of a close network of fibres, looking a little more condensed than the ordinary white matter, with scattered nuclei and a few blood-vessels.

The contiguous portions of the ascending parietal and second frontal convolution present essentially the same change. Although the entire gyrus is involved, the process is chiefly cortical and has resulted in the entire destruction of the gray matter.

I have recently had an opportunity of examining a second specimen at the Elwyn Institution with Dr. Wilmarth, a case of right hemiplegia with idiocy and epilepsy. The brain looked well formed; the meninges were normal. On the posterior part of the first left frontal, about three-quarters of an inch from the fissure of Rolando, there was a depression, over which the membranes appeared normal. The base of the first and the top of the ascending frontal gyrus had an opaque white appearance, contrasting strongly with the surrounding tissue. On section this was seen to be an area of induration passing into the cerebral substance for at least an inch, and cutting, with great resistance. A second opaque-white block existed in the hinder convolution of the quadrilateral lobe. On the left side there were two of these masses, one in the lower occipital convolution, and the other in the supramarginal. These masses were firm, opaque, white in colour, the pia over them not thickened and the contour of the convolutions not destroyed. The examination of the brain has not

yet been completed, but the blocks correspond to the description given by Bourneville and other French writers of the *sclérose tubereuse*.

To the French writers we owe much of our knowledge of cerebral sclerosis. Cazanvieilh,¹ in 1827, described twelve cases under the term *agénèse cérébrale*. Bourneville² has recorded a number of observations from the Bicêtre, while Cotard,³ Jules Simon,⁴ Richardière,⁵ and Delhorme⁶ have more recently written monographs upon the condition. In clinical features the great majority of the cases conform to the description of infantile hemiplegia.

The skull may be flattened on the affected side, and it is not uncommon to find it very broad and prominent above the mastoid processes. In several of the cases the bone on the affected side has been greatly thickened. The dura is usually closely adherent, and osseous plates have been found, which, in Bell's case,⁷ were so extensive that the right hemisphere seemed enclosed in a second bony casing.

The arachnoid is turbid and thickened, and the amount of cerebro-spinal fluid excessive. The pia mater is often so thickened and adherent that, on removal, portions of the cortex come away, leaving a roughened surface, and in these cases the condition suggests a meningo-encephalitis. This localized thickening and adhesion of the pia to the sclerosed convolutions is well illustrated in the specimen just described.

The sclerosis is usually diffuse and involves either an entire hemisphere or a single lobe, or it may be confined to one or two convolutions. In a few cases it has been

¹ 'Archives générales de Méd.,' 1827.

² 'Archives de Neurologie,' 1—5.

³ Loc. cit.

⁴ Loc. cit.

⁵ Loc. cit.

⁶ "Contribution à l'étude de l'atrophie cérébrale infantile," 'Thèse de Paris,' 1882.

⁷ 'Archives générales,' 1831.

in patches—insular. Nodular projections of sclerosed tissue (*sclérose hypertrophique*) may occur over the surface. The affected convolutions are small, of a grayish or gray-yellow color, often with a stippled, pitted surface, to which the pia adheres firmly. In contrast with the neighboring normal gyri, the appearance is very striking and characteristic. The reduction in size of the affected hemisphere may amount to a third of the bulk. In one case the atrophied hemisphere weighed 169 grammes, the normal 653 grammes. The tissue may be a mere shell over a dilated lateral ventricle, as in the cases of Baud¹ and Piorry. In many cases anfractuosités and small cysts have been found in and about the sclerosed tissue. Some of these cysts are evidently the result of old hæmorrhages, as hæmatoidin crystals have been found in the walls. This is particularly the case with cysts in the neighborhood of the basal ganglia. In all of the fifty cases the Rolandic area was involved to a greater or less degree; in some cases affected alone, as in the brain figured; in others, involved in a widespread process. Undoubtedly the motor region, the area of the cortical distribution of the middle cerebral arteries, is most often affected, but this is not always the case.

Thus it is interesting to note the distribution of the lesions in ten specimens of sclerosis of the brain in the Museum of the Elwyn Institution.

1. R. H. Superior parietal lobule, patch $\frac{1}{2} \times \frac{3}{4}$ inch, depressed meninges adherent.

2. R. H. First and second frontal. Occipital. L. H. Occipital lobe almost destroyed. Superior parietal, posterior half. Anterior two-thirds of first frontal; second frontal slightly.

3. R. H. Cuneus. First frontal, extensive; gyrus fornicatus. L. H. Gyrus hippocampus, lingual, and occipital. First frontal slightly.

4. R. H. Occipital lobe, outer aspect; supra-marginal; occipital end of first and second temporals; cuneus

¹ Quoted by Gaudard.

slightly. L. H. Temporal lobe, leaving the first untouched; supra-marginal and the entire angular group.

5. L. H. Gyrus fornicatus almost destroyed. Cyst on the middle of first frontal. R. H. Insula smooth, gyri not evident. Posterior half of first temporal; Sylvian ends of both central gyri. Middle portion of first and second frontal and posterior part of third frontal; gyrus fornicatus.

6. L. H. Normal. R. H. Supra-marginal and angular gyri, encroaching on occipital; posterior third of first, second, and third frontal.

7. Blocks of induration, *sclérose tubéreuse*, scattered over both hemispheres.

8. L. H. Normal. R. H. Temporal, parietal and occipital convolutions uniformly sclerosed; not a normal looking gyrus in any of these lobes. Frontal lobe quite natural looking; occupies more than three-fourths of the hemisphere. Patient was an epileptic; no paralysis.

9. The case given in full. R. H. Normal. L. H. Sclerosis of ascending frontal and part of ascending parietal.

10. R. H. Nodular sclerosis, first frontal and base of ascending frontal. Patch on posterior gyrus of quadri-lateral lobe. L. H. Patch on lower occipital, second on supra-marginal gyrus.

Dilatation of the lateral ventricle on the affected side is frequently mentioned; the basal ganglia may be flattened. In all the recently reported cases a descending degeneration of the pyramidal tract is described. In the cases of extensive atrophy of one hemisphere the opposite hemisphere of the cerebellum has been found smaller. The condition of the vessels has not attracted sufficient attention. In two cases Jendrassik and Marie¹ found important changes in the walls of the cortical arteries to which we shall again refer.

3. *Porencephalus*.—Associated as the condition invariably is with atrophy, of which it is indeed only a final

¹ Loc. cit.

result, to consider it separately is not strictly correct, and yet precision may well yield to the manifest convenience of grouping together for analysis the cases which present this lesion. Porencephalus represents a loss of substance in the form of cavities or cysts, situated at the surface of the brain, either opening into or bounded by the arachnoid, and often passing deeply into the hemisphere, reaching even to the ventricle. It was present in twenty-four of the ninety autopsies in hemiplegia. In eighteen of these cases the onset of the paralysis was noted. In six it was congenital. In eleven the hemiplegia came on in very early life after convulsions. In one it appeared to be the result of a fall at the age of two years. In six of the cases death occurred before the tenth year; in five between the tenth and twentieth year; in five between the twentieth and thirtieth, and in eight after this age. The extent of the defect was very variable, from a few convolutions to half a hemisphere. In all of the cases the motor region was involved to a greater or less extent. Although occurring in other parts of the brain in the great majority of cases the districts supplied by the middle cerebral arteries are affected. It is worthy of note that this condition usually corresponds with definite areas of vascular distribution.

In the recent study of this condition by Audry¹ it is interesting to note that hemiplegia was mentioned in sixty-eight of the 103 cases. In ninety-six cases in which the details were full, the defect was bilateral in thirty-two, the left hemisphere alone affected in thirty-eight, and the right in twenty-six.

DIAGNOSIS.—The more frequent onset with convulsions, the hemiplegic character of the palsy, the absence of rapid wasting of the affected muscles, and the retention of electrical reactions, are features of the early stage of the disease sufficiently well characterized to exclude, in most cases, infantile spinal paralysis. The rigidity of

¹ 'Revue de Médecine,' Nos. 6 and 7, 1888.

the muscles, the increase in the reflexes, the gait, the distribution of the residual palsy, the impaired intellect, and the frequent onset of convulsions clearly characterize the later stages.

There are cases, however, which present difficulties, owing to irregular distribution of the paralysis. Thus the following case of spastic crural monoplegia is probably from its character of cerebral origin.

CASE 25.—James H—, æt. 5 months. I. P., 125. Healthy when born and until two and a half years old, when, for two weeks, he had fever, which was preceded by screaming spells. He seems to have pain in the right leg, and the back seemed painful and stiff. Then rigidity appeared in the left leg. During the attack he was delirious. The arms were never affected. There was no loss of sensation at any time. The condition is now confined to the left leg. The foot can neither be flexed nor extended, and is shorter than the right. Cannot lift the thigh. Can flex the leg slightly. Cannot stand on left leg. There is a difference of one and three-quarters inches in circumference of the legs, and of one and a half inches in that of the thighs. The reflexes are increased.

In the following case there was, at first, right hemiplegia, with the involvement, also, of the left arm, a distribution suggestive of spinal palsy, but the history clearly shows that the origin was cerebral.

CASE 86.—Mary J—, æt. 2½. H., 345. The second of three children; others well. Family history good. Mother fell three days before confinement, and was delivered of the child prematurely at eighth month. Birth normal. About May 30th, when two years old, was peevish and whining for a time, and then had convulsions, in which the mouth and face were twisted to the left, and she moved hands and arms. Both arms were paralyzed; and the right leg after the attack. It was five weeks before she began to use the left hand, and she has never regained power in the right. She began to

use the right foot in about two months. *Status præsens* : Cannot walk, but slides along on her buttocks. Mind is deficient ; cannot talk ; but understands what is said to her. Cranium microcephalic ; biparietal diameter, 4·5 inches ; occipito-frontal, 6 ; circumference, 17½. Slight internal squint. Rigidity of right leg at knee. Right hand is clenched, fingers extended with difficulty ; hand seems slightly smaller than the left ; no difference in forearms. Right hand gets cold and blue at times.

At the Elwyn Institution there is a remarkable case which must be regarded, I think, as one of post-hemiplegic epilepsy. All trace of the hemiplegia has disappeared, but there remains slight wasting, increase of the patellar reflex, and occasional attacks of Jacksonian epilepsy, to indicate the nature of the original trouble. As the fits became general, and were aggravated at each menstrual period, oöphorectomy was performed, with relief at first, but, as might be expected, the attacks have returned.

CASE 23 (Elwyn series).—Annie S—, æt. 23. Inmate of institution for one year. Mother is in an insane asylum. When between three and four years old she fell and, for a long time after, could not walk ; weakness of the left side. In one history of this case it is stated that the paralysis came on after scarlet fever. Had spasms at the age of twelve. Always began in arm, and for a long time were in the arm alone. Had the spasms worse at menstrual period, usually three or four, either just before or just after the period. Oöphorectomy was performed two years ago, February, 1886. Better for a year. Is a well-nourished girl ; gives a good account of herself. Left arm is smaller than right. Left forearm, 9 inches ; right 9½. Wrist : left 6½ and right, 7. No spasm, no rigidity ; movements perfect. Left leg smaller than right. Calf, 12⅞. Right calf, 14⅓. Foot ; length : left, 8⅓ ; right, 9. Left knee-jerk +. No ankle-clonus ; no heart lesion. The fit begins every time in the left arm, the fingers are drawn up and the arm gets stiff.

She has then to lie down on the sofa, and they become general; sleeps after them; has often vomiting. Has not had an attack since February, 1888. At first, while in the institution, they recurred for a number of months with regularity.

In certain cases of *cerebral tumour* the symptoms are those of spastic hemiplegia. Seeligmüller¹ records such an instance of tubercle of the meninges, beginning with convulsions and fever, with left hemiplegia and contractures in hand and foot; and I have described a case of glioma² in a girl of thirteen years, who had spastic hemiplegia and Jacksonian epilepsy for some years.

Paralysis of the face and of the upper extremity may result from the application of the forceps; the latter more commonly from the manipulations of the accoucheur. These *obstetrical paralyses*, as Duchenne³ called them, could rarely be confounded with those due to cerebral causes. They are fully considered by Nadaud,⁴ and Budin⁵ had recently given an excellent description of the brachial palsy.

PROGNOSIS.—Parents are naturally intensely anxious as to the prospects of a child attacked with hemiplegia. As a rule, the younger the subject the greater the liability to serious and permanent damage. The nature of the lesion doubtless has an important influence, vascular obstruction being more favorable than meningo-encephalitis, and the latter than tumour, but the diagnosis of the exact condition is by no means always easy. Of the bodily defect, about which the greatest uneasiness is displayed, we can almost invariably predict great improvement, particularly of the leg. Perfect recovery of the arm is rare, of the face more frequent. The development of post-hemiplegic movements is a bad omen, as such cases are more likely

¹ 'Jahrbuch für Kinderheilkunde,' Bd. xiii.

² 'American Journal of the Med. Sciences,' 1885.

³ 'Traité de l'électrization localisée,' 3rd edition.

⁴ 'Des Paralysies Obstétricales des Nouveau-nés,' Paris, 1872.

⁵ 'Le Bulletin Médical,' No. 20, 1888.

to have epilepsy, which constitutes one of the most serious sequences of the disease. Not only are the attacks dangerous in themselves, but they undoubtedly tend to aggravate existing mental defects. They may not develop for some years after the onset of the paralysis and may be deferred till the period of puberty. More distressing still to the relatives is the enfeebled mental state which so often follows infantile hemiplegia. As the records of the Pennsylvania Institution for Feeble-minded Children at Elwyn show, the percentage of the cases due to this cause is by no means small. Training does much for them, but for too many the outlook is not hopeful, and as Merklin states, even when in childhood they escape imbecility, they are very liable in adult life to become the subjects of psychoses. *Quo ad* life the prognosis is good, as shown in the post-mortem reports of ninety cases in a large proportion of which the age at death was over twenty years.

CHAPTER III.

BILATERAL SPASTIC HEMIPLEGIA.

SYNONYMS.—Spastic rigidity of the newborn (Little). Tonic contraction of extremities. Essential contraction. Spastic rigidity. Permananter-kinder tetanus (Stromeyer). Spastic paralysis of children (Adams). Spastic diplegia (Gee). Spasme musculaire idiopathique (Delpech).

In infantile hemiplegia a great majority of the cases occur within the first three years of life, and in only a limited number is the condition congenital, either the result of intra-uterine disease or of accident during parturition. In bilateral hemiplegia and in paraplegia the reverse holds good; in a large proportion of the cases the trouble dates from birth, and is the result of injury to the child during its passage into the world. Hence the appropriateness of the term birth palsies, applied to these cases by Gowers. Strictly speaking, these cases should be considered together, as they depend on essentially similar conditions, and we may find the arms so slightly affected that it is difficult to say whether the case is one of diplegia or paraplegia; but there is a sufficient number of clearly defined cases in each group to make a division advisable, and there are questions relating to the spastic paraplegia of children which deserve separate consideration.

To the orthopædic surgeons we owe the greater part of our knowledge of these cases. Heine¹ understood them thoroughly, and to him, I think, belongs the credit of first recognizing their cerebral origin, and separating them

¹ 'Spinale Kinderlähmung,' zweite Auflage, 1860.

from the ordinary infantile paralysis. He clearly distinguished cases of *hemiplegia cerebialis spastica* and *paraplegia cerebialis spastica*, using these expressive names which have since been employed with minor modifications by Benedickt, Bernhardt and others. At page 163 of his monograph is also to be found, perhaps the first, certainly a most accurate, account of post-hemiplegic movements.

Little, the well-known London orthopædic surgeon, has contributed more than anyone to the subject, and to him we owe, in great part, the accurate knowledge of the relation of the cases to abnormal parturition. His paper in vol. iii of the London Obstetrical Society's 'Transactions,' 1862, contains an immense amount of material. The clinical description which he gives at pages 301—303 has not been excelled. In France the cases of spastic rigidity are sometimes called Little's disease (*maladie de Little*). The writings of Delpech,¹ Stromeyer² and Adams³ particularly of the latter, to whom we owe the name spastic paralysis of children, contain careful descriptions; and more recently Rupprecht⁴ has considered the subject from a surgical standpoint.

SYMPTOMS.—Bilateral hemiplegia is characterized by a spastic condition of the extremities, dating from or shortly succeeding birth; occasionally following the specific fevers or an attack of convulsions. The legs are more involved than the arms; there is no wasting; no disturbance of sensation; the reflexes are increased. The mental condition is profoundly disturbed; the patients are usually imbeciles or idiots, helpless in mind and body. Ataxic and athetoid movements of the most exaggerated kind may occur.

¹ 'Orthomorphie,' Paris, 1828.

² Stromeyer, 'Handbuch der Chirurgie,' Bd. ii.

³ Adams, 'Club-Foot,' London, 1866.

⁴ Rupprecht, "Ueber angeborne spastische gliederstarre und spastische contracturen," 'Volkmann's klin. Vorträge,' No. 198.

Anatomically there is bilateral sclerosis or porencephalous defect of motor areas of the cortex cerebri.

CASE 1.—Anna S—, æt. 3½. First child ; no instruments used in labor ; full term. An “inward convulsion” on tenth day ; lay for three days apparently dead. First teeth came at sixth month. Child has not progressed well. Head seventeen and a half inches in circumference.

February 29th, 1888.—Aged now six years ; says only “mamma,” “papa” and “no.” Strabismus, no nystagmus. Has thriven and grown, understands everything. Head long and narrow ; biparietal diameter five inches ; occipito-frontal six and a quarter inches ; circumference eighteen and a half inches. Face blank, but laughs. Conjunctivitis ; teeth bad. Uses hands to eat ; arms stiff at elbows. Legs stiff in extension. Well nourished. When cries gets very rigid. Pes equino-varus. Feet cross when attempts to stand ; cannot walk ; knee-jerk +, no clonus.

CASE 2.—Amelia P—, æt. 14 months. M. 5, 423. Born at seven months ; no instrument ; reason assigned for miscarriage carrying heavy weight. First child. Great weakness noticed at birth, but nothing else ; child very small ; no spasms ; has never had any serious illness ; has never cut any teeth ; never walked ; never talked, except for the last two months to say “papa” and “mamma.” Convergent strabismus. Never has had any skin eruption. Keeps the legs crossed. Recognizes the parents ; does not fix attention ; does not sit up well ; wobbles the head about. Knee-jerk + +. Plantar reflex +. Head microcephalic, symmetrical ; fontanelles closed ; circumference of head seventeen and a half inches ; occipito-frontal diameter six inches ; biparietal diameter five and a half inches ; bridge of nose sunken. Arms and legs stiff, moves them with freedom ; takes things with the left hand, does not grasp well ; right arm especially stiff at shoulder and elbow ; while under observation legs stiffened in extension ; in the interval the legs can be

extended and flexed easily; stands fairly well when supported.

CASE 3.—Everett A. P—, æt. 5. Parents alive and healthy; no nervous diseases in family. This child is the oldest of three. Born at seven months; no instruments; small, very weak, jaundiced. Noticed stiffness when only a week or two old, in legs and arms; has never walked. Does not know his letters. Never had a spasm; had scarlet fever. Hands have always been free from spasm; elbows stiffer sometimes than at others. Temper not very good. *Status præsens*: Legs: color, temperature, nutrition good. Knees, ankles and hips stiff; cannot walk, when supported stands on toes with legs crossed. Spasm in facial muscles at times. Knee-jerk+. Can use hands well; elbows stiff. Cremasteric and abdominal reflexes present. Spine straight. Phimosis; teeth fair. No nystagmus. Bright, talks well. Head—biparietal diameter five and a half inches; occipito-frontal seven inches; circumference nineteen and three-quarter inches. In bed sits with spine doubled, cannot sit up straight.

CASE 4.—Florence S—, æt. 4. M., 5, 137. No forceps used. No trouble during pregnancy. Five older children. Father and mother healthy. Nothing special about child at birth; nursed well. Spasms on the second day. Has not walked or crept. Does not talk, understands what is said. Is well-nourished, fairly well-grown child; looks bright and is fairly intelligent, though expression is spoilt by the constant dribbling from the open mouth. Head wobbles from side to side. Head measures nineteen and a half inches in circumference, and is symmetrical. Sutures closed, and a little ridged in their course. Arms not well developed, and are stiff, but can be readily flexed at elbows, wrists and fingers. Gets so stiff at times that she can be lifted without bending. Arms stiffen in extension. Moderate grasping power. Knees and thighs can be flexed, abducted and adducted. Like arms, they stiffen and get hard. Cannot

walk. Plantar reflexes not marked. No ankle-clonus. Knee-jerk+. Teeth much decayed. No convulsions now. Sleeps well, but will not sleep in the dark, and awakens at once if light is removed.

February 4th, 1888.—Will be six years old in August. Is small for her age. Does not get so stiff now, only once or twice a day, not, as before, a dozen times. Very characteristic posture of legs—feet extended and legs crossed. Stands on toes, generally on right foot, with the left leg diagonally across the right. Holds her head better; is not cross. Hands get stiff, and fingers separate and extend when she attempts to take anything.

CASE 5.—Jennie S—, æt. 10. M., 7, 177. Labor difficult. Head much flattened in birth, and child was unconscious for half an hour. First child. Second died at four years from accident. Mother thinks child always used arms and legs with difficulty. Never learned to walk or use the hands. Dentition natural. Intellect deficient, yet understands ordinary conversation. Has not learned anything. Knee-jerk++; muscles rigid; arms and legs almost contracted. Color pale, appetite good, sleeps well. Bowels regular. Heart normal. Lower jaw retreats. Occipito-frontal diameter, six and a half inches; occipito-mental diameter, eight and a quarter inches; biparietal diameter, five inches.

CASE 6.—Lydia B—, æt. 2 years and 3 months. M., 185. Elder of two children, other healthy. Born without instruments. Head slightly microcephalic. Biparietal diameter, four and three-quarter inches; occipito-frontal, five and a half inches. Fontanelles closed. Forehead prominent in central portion. Internal strabismus. Mouth open, no dribbling. Intelligence poor, speaks little, smiles, and looks intelligent. Dentition began at six months, last tooth is now being cut. Arms stiff, especially left. For a time kept left hand closed. Legs stiff, slight extension of feet. Tendency to talipes equinus, legs flexed with difficulty, sits with legs crossed. Never has used legs properly, they are thin and cold. Electric

examination shows the quantitative change, but no reaction of degeneration. She had a spasm soon after first visit ; tendon-reflexes increased.

CASE 7.—Nellie M—, æt. 6. M., 5, 291. Mother died of phthisis ; one child died at eleven months, of convulsions, while teething. This child is one of twins, the other born dead. Some defect noticed at birth ; teething natural. Never had convulsions. Never has been able to sit up or walk ; has to be fed. Right hand weak and small, can take some things in it. Both shoulders stiff, a little pressure relaxes them. Thighs are crossed. Legs have clasp-knife rigidity ; left leg crosses right ; this position is nearly constant. Legs can, however, be placed side by side ; right leg decidedly larger than left. Foot strongly extended on leg ; ankle-joint cannot be flexed. Knee-jerk++ ; no ankle-clonus. Expression idiotic ; speaks no word ; temper bad ; mouth open ; constant dribbling ; does not cry much. Head microcephalic, supra-orbital arches marked. Face prognathous. Forehead low, narrow ; does not support head, but it wobbles from side to side. Circumference of head, seventeen and a half inches. Chest rickety, costal margins everted ; sternum depressed ; antero-posterior and lateral spinal curvature. Convergent strabismus, nystagmus.

CASE 8.—Harry B—, æt. 1. M. 5, 168. One brother said to have some brain trouble, one died in a convulsion at the age of three years, one died of congestion of brain. Two other healthy children. Fourth child ; labor natural. Was backward about walking. Since summer of 1885 has twitched hands and feet. Head symmetrical, measures nineteen inches ; expression bright and intelligent. Temper good. Makes no sounds, seldom cries. Uses his hands well ; at times clinches hands and folds them over heart, and they get stiff. Eyes turned a little. Moves feet with a slight spasmodic movement. Legs cross when at rest, and when he is lifted they get stiff.

CASE 9.—Wm. R—, æt. 2. M., 4, 33. Family history

good. Mother had a severe fall five months before child was born. Never has had any children's diseases. Is moderately well nourished. Special senses perfect ; dull, stupid, never says anything except "mamma." During last year has had frequent attacks, in which face grows at first scarlet, then purple, and finally white. Attacks last from two to three minutes, during which he works mouth and twists lips. There is entire loss of consciousness. Does not bite tongue or froth at mouth ; attacks occur in daytime ; sleeps after them. Independently of these attacks during the day he has repeated spasms of the muscles at the back of the neck, and the head is drawn into complete extension. There is a constant spasm of the left thumb, which is drawn across the palm of the hand. Arms stiff. Legs tend to cross when he is held erect, but can be spread apart by force ; great difficulty in flexing the legs when they are in spasm. Cannot stand alone, and has never walked more than two or three steps. Uses hands but little, holds objects in an awkward manner, and cannot feed himself. Still nurses, but takes other food. Slight nystagmus. Dribbles constantly. Sleeps well, but starts at sounds ; is constipated.

CASE 10.—Morris C—, æt. 3. M., 4, 122. First child ; labor lasted eleven hours ; instrumental. Father thinks mother was frightened when half way through pregnancy. Child's body was black when born. He cried continuously for one week after birth. When he began to move, parents noticed his movements were awkward. Cannot sit up. Has no fits now. Is said to have had them last fall. Appetite and sleep good ; shows signs of intelligence. Eyes squint internally. February 15th, 1888.—Returns to-day. Is now eight years old. General health has been good ; no convulsions ; has not been able to sit up, nor walk ; lays on stomach in cot-bed most of the time ; cries very little, temper good ; can scarcely talk, says a few words—*e. g.*, "home," "all right," "hello." Looks fairly bright and intelligent, and understands what is said. Head not quite symmetrical ; left parietal and

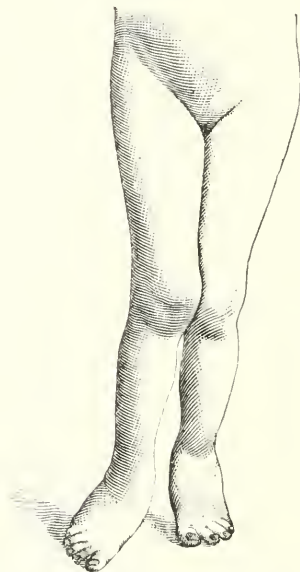
left frontal eminences most marked. Biparietal diameter, five inches; occipito-frontal diameter, seven and one-quarter inches; circumference of head, nineteen and a half inches. Eyes bright, clear, slight convergent strabismus; pupils medium size, react to light, vision seems good. Makes irregular movements of lower muscles of face; opens mouth, and utters unintelligible sounds. Has difficulty in supporting head, which wobbles from side to side. There does not appear to be distinct spasm of the neck muscles. Back much turned to right. No wasting; body is thin. Hands are held closed, opens them sometimes; thumb inverted, and fingers usually closed over it; is unable to grasp anything. There are constant, large, irregular movements of arms; distinct spasm in arm; very difficult to extend forearm. Right thigh constantly drawn up in semi-flexed position; leg flexed on thigh; left leg extended, foot inverted, great stiffness; muscles of legs fully developed, but firm. Knee-jerk +; no ankle-clonus. Teeth good, getting second teeth.

CASE 11.—Thos. McG—, æt. 4. M. 5, 195. One of three children. Born naturally, and seemed healthy. Measles at three months, followed by hydrocephalus; head continued to increase in size till one year old; circumference twenty-three and a half inches. Dentition normal. Talked at an early age and seemed quite bright. Never walked, but can kick legs; legs spastic on standing, toes extended, muscles tight, feet crossed, legs relaxed when at rest. Knee-jerk + +. Hands shake and arms are stiff.

CASE 12.—Elsie J—, æt. $1\frac{1}{3}$. M., 5, 216. Roxborough. Youngest of two children, the other perfectly well. Labor natural and not difficult. Never had convulsions; no definite onset. Head microcephalic, measures seventeen and a quarter inches in circumference; furrow in temporal region; sutures closed for some time; transverse diameter above ears four and a half inches; occipito-frontal, six inches. Dentition retarded. Has upper incisors and

lower central. Seems bright and notices objects. No nystagmus. Fairly well nourished. Arms stiff at elbows and shoulders; knee-jerk +. Legs were noticed to be stiff soon after birth; not well developed; never walked; on standing rests on toes; (Fig. 3) knife-clasp rigidity;

FIG. 3.



Position of child when supported.

strong adduction; sometimes crosses legs; some eversion of feet; attempts to walk; seems to have more power on left side. January 31st, 1887.—Supposed to have had spasms about a year ago; was three years old last December. Arms and legs remain stiff. Child has not developed mentally.

CASE 13.—Willie L—, æt. 6. One child stillborn, one, aged two weeks, died of marasmus. Born with instruments; had fits as soon as born, lasting for three days; moved arms and legs, but not so well as other children;

never able to sit up till one year old ; began to talk at two years ; sat up and held up head at eighteen months ; right hand contracted till second year ; dragged the right hand in creeping ; stood at three and a half years ; cannot stand now without support. Intellect bright ; speech affected, has difficulty in pronouncing words. Fits again two years ago, after improper food. Healthy but small. Back straight ; walks on toes ; legs small below knees ; skin and tissues adherent ; contraction of tendo Achillis, which can easily be overcome. Legs cold ; in walking toes of left foot turn in ; left hand rather the larger ; both arms stiff.

CASE 14.—Fred H—, æt. 3. M. 7, 58. Parents healthy, five other children, all well. Looks bright ; understands everything. Born at seven months ; no instruments ; nursed by mother. Has never sat up, crawled, or walked ; is now just beginning to say a few words. Restless and irritable ; constantly throws himself backward. Hands and arms contracted and stiff, and in constant motion ; makes an offer to take things, but can scarcely take anything to his mouth as the arms are so stiff, left more than right ; feet are at times everted, then the toes are flexed and extended. Knee-jerk difficult to obtain ; no ankle-clonus ; spasm gets less when once overcome by motion ; teeth good ; no nystagmus. Biparietal diameter four and three-quarter inches ; occipito-frontal seven inches ; occipital region very prominent ; circumference nineteen and a quarter inches. He has been ill lately with fever ; is now very pale ; at times gets very stiff in arms and legs ; hands and fingers extend and get rigid ; uses left hand most ; can scarcely grasp an object. Has had two slight convulsions, limited almost entirely to face and mouth, first last autumn (1886), second in March, 1887.

CASE 15.—Ralph W—, æt. 3 years and 6 months. In Pennsylvania Institute for Feeble-minded Children. Idiot ; no history ; unable to walk or talk. Head microcephalic, circumference sixteen and a half inches, with the hair ;

no nystagmus; does not dribble; teeth good; arms rigid, and become more so on the slightest touch; irregular movements in fingers, but how much voluntary and how much involuntary, it is difficult to say; legs in extreme spasm, slight equino-varus position; the feet can scarcely be moved, so firm is the extensor spasm; the whole pelvis moves on attempting to flex the thighs.

A class of cases belonging to this division of bilateral hemiplegia is characterized by *spasm and disordered movement*. They are described in literature as *chorea spastica and double athetosis*. The cases I refer to are simply spastic diplegias, plus post-hemiplegic disorders of movement. The history is the same as in ordinary cases; the trouble has persisted from birth or shortly after, and there is a condition of feeble-mindedness or idiocy, though in some instances the intelligence is fair. Very often, too, there has been a difficult labor.

Of the chorea spastica, the following are probably illustrations:

CASE 16.—Mary M—, æt. 4. M. 4, 429. Breech presentation, delay at the head, was six hours before she was resuscitated. Began to talk at two years; never walked; almost from birth she has had peculiar movements of hands and arms; the thumbs are turned in and there is constant irregular motion of the arms and hands, with stiffness, which is made worse when she attempts to control it; it is like a chorea. There is also some incoördination of the head. She is well nourished; no wasting. Coördination of legs good; but she does not walk.

CASE 17.—Nellie P—, æt. 9. I. P. b., 55. Parents healthy, five children dead, of seven. Seven years ago had fits while teething, had fits constantly for twenty-one days; for nine months had seven to nine per diem; in very weak health when fits ceased. *Present state*: Speech hesitating; memory not affected; unable to stand, sit, feed herself, or assist herself in any way; can move every

muscle in the body, but with an irregular movement which prevents her using any group of muscles; the movement is choreoid; in attempting to grasp an object the fingers are thrown out in a stiff, spasmodic, and irregular manner, and she is unable to close them over the object.

I do not mean to infer that all cases of so-called congenital chorea come under this designation; there are instances without spastic rigidity, as the case reported by Dr. Sinkler from this Infirmary.¹ Certain of these cases of congenital chorea have also had definite athetoid movements.²

There are several reports of children in the Infirmary records with the diagnosis of multiple sclerosis, which in many respects resemble these cases, and it would doubtless often be difficult to make a differential diagnosis.

Bilateral athetosis is not very uncommon; an illustration may be found in almost every almshouse or home for incurables. It is one of the most distressing of all maladies to witness, and is usually associated with imbecility.

The following cases illustrate the combination of spasm with disordered movement characteristic of this condition:

CASE 18.—William B—, æt. 30. In the Elwyn Institution for eight and a half years. History: Had jaundice when eleven days old, after which the paralysis occurred. *Status præsens*: Intelligent looking; head well formed. Does not speak, but utters a loud, deep-toned sound when he is pleased. Sits up, but in a sloping position. Cannot stand. Continual grimaces, caused by irregular movements of the lower face muscles. Head is turned forcibly from side to side and the mouth drawn and hideously distorted. Arms very stiff, not wasted; are quiet at times, but every few minutes the most irregular

¹ 'System of Medicine,' edited by Pepper, vol. v, Philad., 1886.

² Rau: 'Neurologisches Centralblatt,' 1887.

movements ; the arms and forearms stiffen in extension, the hand flexed, and the fingers in rapid, continuous spasm. At times is quite quiet and can even feed himself. Sits usually with his wrists strongly flexed on the bench, as if helping to support himself. The motions of the fingers are typically athetoid ; those of one hand will be flexed on the palm while the others are in active extension. As he feeds himself the spasm is very great, and it is with much effort that the mouth and hand can be made to meet. The index finger may be strongly flexed while the middle is in extreme extension. The legs are stiff, strongly adducted. The feet are in extension in equinovarus position. The knee-jerk is obtained with difficulty. He is good-tempered and smiles ; knows the attendants and makes signs as to his wants.

CASE 19.—Laura C—, æt. 21. The fifth child. A hard labor, but no instruments used. When born there was “no sign of life in her,” and for an hour she was blue. The mother is a large well-built woman ; the family history is excellent. At six months the child had whooping-cough ; seemed weak before this, but after the attack grew much worse ; could not sit up and could not help herself like other children. From infancy she has had irregular movements of the arms and legs, with stiffness. Learned to talk late ; is intelligent and good-tempered. She was brought to the Infirmary when eleven years old. When seen recently at home she presented the following condition : a medium-sized girl, pale, but with an intelligent face. Sits in a chair supported by cushions. She answers questions in an interrupted, somewhat high-pitched voice, a little difficult to understand at first. Mentally is quite bright ; appreciates her condition, and said she was a “little Job.” Likes to be read to and to play with the children. She has never walked, and is quite helpless on account of the extraordinary rigidity and irregular movements of the extremities, which are excited by emotion or by any attempts at voluntary efforts. The facial muscles move spasmodically as she speaks. When

pleased she laughs in a loud, rough manner, with the mouth widely opened, the jaw strongly depressed, so that the uvula and palate are freely exposed. The arms are well nourished and are held in strong extensor spasm; the left is rotated inward and rigid; the forearm is so strongly extended that there is almost an anterior dislocation of the elbow-joint. At the same time there is extreme rotation of the radius and the hand, the fingers of which are clinched so tightly that it is impossible to separate them. The right arm is less strongly contracted, and with it she can make attempts to grasp objects. The spasm relaxes every few moments and the limbs assume new attitudes. The fingers relax and close, but without that continuous, orderly spasm seen in typical athetosis. The shoulder and trunk muscles are also affected, and their irregular contraction moves the trunk about from side to side. The legs are strongly extended, the feet in the equino-varus position. There is not much movement, but on testing the reflexes there were sudden spasmodic jerkings, and at times the knees are drawn up. The muscles in spasm have an iron-like rigidity, and it is almost impossible to bend the limbs. When not excited she is much quieter and the muscles relax; but the slightest exertion brings on the spasms. She can sometimes, with the left hand, pick up objects, and even carry a biscuit to her mouth, but she is quite unable to feed or help herself. The arm- and leg-reflexes are increased; the ankle-clonus is readily obtained. Sensation perfect; the most extreme spasm gives no pain. There are no trophic changes. Her appetite is good and she sleeps well, the contractions disappearing completely. The bodily functions are well performed.

Recently a young man came to the Infirmary whose case forms an interesting link between the common forms of spastic diplegia and those associated with disordered movements:

CASE 20.—E. A—, æt. 21. Eldest child. Difficult labor. The mother died in childbed with third child.

Very limp and feeble as an infant. Had two fits in early life and one at age of five years. Did not walk until the ninth year as "the cords at the back of his legs were too tight." Has been bright and intelligent, but did not learn to read until late. *Status præsens*: Intelligent looking: head well formed: circumference twenty-two and a half inches. The muscles of the face move irregularly, and the lips are drawn up. The speech is imperfect; articulates with decided effort, and there are clonic contractions of the facial muscles. He can stand and get up alone; walks with difficulty; steps are short, and the gait is very stiff. Leg muscles are not well developed. Left calf measures eleven, right eleven and a half inches. On attempting flexions legs get very stiff. Arms fairly well nourished. Supinators are small. No movements when at rest, but when excited or when attempting any action, the arms get stiff and the motions are very slow. He can dress himself, using the fingers in a stiff clumsy manner. The reflexes are increased; ankle-clonus present. Sensation perfect. Special senses unimpaired.

In spastic diplegia some of the patients are able to walk; in all the hands are used awkwardly, or not at all. The legs are most affected, usually extended, the feet crossed and in the pes equinus or equino-varus position. The thighs are often strongly adducted—the so-called clasp-knife rigidity. When the child sits the legs cross, and, if supported, there is the characteristic attitude of infantile spastic paraplegia, the feet crossed and the body supported on the toes. In Case 10 the right thigh was drawn up; sometimes the legs are partially flexed, but, as a rule, the extension position is maintained. The stiffness is in some cases constant, while in others it varies greatly and is increased when the child cries, or in attempts to move. It may be more marked on one side than on the other. The whole body may at times become rigid, and, as Little remarks, is turned "all of a piece" on the lap. The arms are usually flexed, and the stiffness

is at once apparent on attempting to extend them. It may be difficult or impossible to raise the arms or to abduct them. The hands may be clenched and the fingers strongly flexed, but it is rare to see the extreme spasm which is so common in hemiplegia; and, as the reports indicate, a majority of the children could use the hands, though awkwardly; while in Case 3 the movements were almost natural. There may be, as in Case 10, large, irregular movements of the hands.

Spasm of the muscles of the face or tongue was rarely noted. In Case 10 there were irregular movements of the facial muscles; and a condition of rigidity was at times present in the facial muscles. The spasm is not always fully relaxed during sleep, but disappears when the patient is fully etherized. The back and neck muscles are weak, and the child is rarely able to sit up alone. The spine in some cases seems to have remarkable flexibility, and mothers have used more than once the expression that the child was as "limp as a rag." So helpless, indeed, is the condition of many cases that unless in bed they must be in the lap. The feeble neck muscles are unable to support the head, which rolls from side to side or sinks on to the chest. The muscles were firm and hard, not often marked wasting.

In no case was sensation impaired.

The reflexes were increased in all these cases, particularly the knee-jerk. The ankle-clonus cannot, as a rule, be obtained.

The electrical conditions in the cases tested were unchanged.

With the exception of Cases 3, 19, and 20, the children were either idiots, with a glimmer of intelligence, or imbeciles. The facial expression usually indicated the mental deficiency. The open mouth, constantly dribbling saliva, and lolling tongue were present in the majority of the cases. Only two of the children could speak plainly, thirteen could not talk at all, and six spoke with difficulty. With the exception of two or three cases,

they all seemed to be able to understand, more or less, when spoken to by their mothers. Irritability of temper was complained of very much.

Microcephalus is a very common condition with asymmetry, and in several of the cases the head was very broad above and behind the ears.

In two cases nystagmus existed, and in three strabismus.

As is so common in imbecile children, the teeth were defective, a condition to which Dr. Alice Sollier has recently devoted a special monograph.¹

Only two children had epilepsy, Case 9, subject to attacks of *petit mal*, and Case 13, which had two spasms in which the face twitched, and the child seemed to lose consciousness. Cases 1 and 4 had had convulsions after birth, and Case 10 had fits at two years of age.

Of these twenty cases the youngest at the time of application was one year and the oldest ten. Six were first children; three were born at seven months; in three cases the labor was prolonged, in two of which forceps were applied; one was a breech case. In eight cases the labor is stated to have been natural.

In ten of the cases the condition was probably congenital, as there was no definite onset, and the stiffness was noted early. In Case 1 there were convulsions on the tenth day, and the child was unconscious for three days. In Case 6 the head was much flattened by the forceps, and the child could not be roused for half an hour. In Case 10, also, the child was delivered with forceps, was asphyxiated when born, and cried for a week. In Case 11 the child had measles at three months, which was followed by hydrocephalus and the gradual development of a spastic condition.

MORBID ANATOMY.—I have been able to collect the reports of sixteen autopsies in cases of bilateral spastic

¹ 'De l'état de la Dentition chez les Enfants Idiots et Arrières,' Paris, 1887.

hemiplegia in children; the youngest was two years old, the oldest thirty. The anatomical condition in these cases was as follows: Case 1. F—, æt. 5 (Kundrat¹). Bilateral porencephalus, motor regions. Case 2 (Henoch²). M—, æt. 6. Atrophy, frontal convolutions. Case 3 (Heubner³). Æt. 2½. Atrophy of left central and right parietal convolutions. Case 4 (Ross⁴). F—, æt. 2½. Bilateral porencephalus. Case 5 (McNutt⁵). F—, æt. 2½. Bilateral atrophy, central convolutions. Case 6 (Richardière⁶). F—, æt. 2½. Sclerosis of temporo-occipital and parietal gyri on both sides. Case 7 (Isambert and Robin⁷). Æt. 2. General cortical sclerosis of both hemispheres. Case 8 (Bourneville⁸). Æt. 10. Extreme sclerotic atrophy in both hemispheres. Case 9 (Bourneville⁸). Æt. 9. Bilateral atrophy of convolutions, particularly the central gyri. Case 10 (Blanchez⁹). F—, æt. 5. Atrophy of posterior lobes of both hemispheres. Case 11 (Simon¹⁰). Æt. 2½. Sclerosis of central convolutions. Case 12 (Bourneville⁸). Æt. 5½. Foci of sclerosis in frontal and temporal lobes. Case 13 (Ashby¹¹). Æt. 22 months. General atrophy; surface of hemispheres smooth. Case 14 (Moore¹²). Æt. 5. General cortical sclerosis. Case 15 (Gee¹³). F—, æt. 11. General cortical sclerosis. Case 16 (Mierzejewsky¹⁴). Æt. 30. Double porencephalus.

¹ Kundrat, 'Die Porencephalie,' 1882.

² Henoch, 'Lectures on Diseases of Children,' American edition, 1882.

³ Heubner, 'Berliner klinische Wochenschrift,' 1882.

⁴ Ross, 'Brain,' vol. v.

⁵ McNutt, loc. cit.

⁶ Richardière, loc. cit.

⁷ Isambert and Robin, quoted by Wuillamier, loc. cit.

⁸ Bourneville, quoted by Wuillamier.

⁹ Blanchez, quoted by Wuillamier.

¹⁰ Simon, loc. cit.

¹¹ Ashby, 'British Medical Journal,' 1886, i.

¹² Moore, 'St. Bartholomew's Hospital Reports,' xv.

¹³ Gee, 'St. Bartholomew's Hospital Reports,' xvi.

¹⁴ Mierzejewsky, 'Archives de Neurologie,' tome i.

A more detailed account of Dr. Sarah J. McNutt's case will illustrate the condition which exists in the majority of these patients. The child, two and one-half years old, had been delivered with instruments and had convulsions during the first nine days of its life, and for a long time did not seem to have any muscular power. When first observed there was paresis with rigidity of all the extremities, and the child was defective mentally. Death occurred from gastro-intestinal catarrh. The brain was studied by Dr. William H. Welch, who has given a very full description of the coarse and microscopic appearances. There was atrophy in each hemisphere of the paracentral lobule, of the central convolutions and of the roots of the three frontal convolutions. Microscopically the cortex of the affected convolutions was replaced by a finely fibrillated tissue, rich in nuclei and without ganglion-cells and nerve-fibres. There was typical bilateral secondary degeneration of the pyramidal tracts in the pons and medulla and cord. In the pons most of the bundles of the longitudinal fibres were degenerated, in the medulla the sclerosis was confined to the anterior pyramids, and in the cord the degeneration involved the direct and pyramidal fasciculi on both sides. The ganglion-cells of the anterior horns were normal in number, size, and general appearance.

Destruction of the motor centres of the cortex is, then, the essential lesion in bilateral spastic hemiplegia. Diffuse atrophic sclerosis is the most common condition; a patchy sclerosis has been found in some cases; porencephalus in others, while in Ashby's case there appears to have been arrest of development, as the surface of the hemispheres was smooth and sclerotic. Descending degeneration has been found in the pyramidal tracts in the cases of McNutt, Jules Simon, and Ashby. In the majority of the cases there was no report as to the condition of the cord. In Ross's case the cord did not show any changes.

Voisin¹, in 1884, communicated to the Paris Academy

¹ 'Bulletin de l'Académie de Médecine,' 1884.

of Medicine a note on the morbid anatomy of five cases of this kind. No details were given, merely the statement that the condition was caused by arrest of development and atrophy of the central gyri. I have not been able to find a full report of his paper.

CHAPTER IV.

SPASTIC PARAPLEGIA.

SYNONYMS.—Paraplegia cerebialis spastica (Heine). Tetanoid pseudo-paraplegia (Seguin). Spastic spinal paralysis (Erb). Tabes dorsalis spasmodique (Charcot).

Spastic paralysis of the legs in children is a common affection, and yet it is only within the past few years that the subject has attracted much attention from writers on diseases of the nervous system. The orthopædic surgeons have for years past described and figured cases which in reality form no inconsiderable quota of the patients at their clinics. Heine, as early as in 1840, gave an admirable account of it, and expresses the modern conception of the disease in the name which he applies—*paraplegia cerebialis spastica*. Delpech, Stromeyer, Adams, and more particularly Little, describe it in their works already referred to. Erb¹ and Seeligmüller,² in Germany, and Gee,³ in London, brought the subject to the notice of physicians, and the first-named author described the cases with those of spastic paraplegia of adults. Ross,⁴ Hadden,⁵ Gowers,⁶ d'Heilly,⁷ and Gilbert⁸ have more recently dealt with the question, and the disease is now

¹ Erb, 'Virchow's Archiv,' Bd. lxx.

² Seeligmüller, 'Jahrbuch für Kinderheilkunde,' Bd. xiii.

³ Gee, 'St. Bartholomew's Hospital Reports,' vol. xiii.

⁴ Ross, 'Brain,' vol. v.

⁵ Hadden, 'Brain,' vol. vi.

⁶ 'Diseases of the Nervous System,' vol. i.

⁷ 'Revue men. des maladies de l'enfance,' 1883-84.

⁸ 'Revue médicale de la Suisse romande,' 1887.

usually assigned a place among the cerebral palsies of children.

SYMPTOMS.—The general features of the disease may be thus defined : Spastic paralysis of the lower extremities dating from birth, or coming on within the first years of life ; absence of wasting ; a condition of talipes equinus or equino-varus ; adductor spasm, producing the “ clasp-knife rigidity ;” the gait stiff, the patient walking on the toes, or there may be cross-legged progression. The intellect is usually impaired, though not, as a rule, so profoundly affected as in bilateral hemiplegia.

CASE 1.—Samuel B—, æt. 14. Parents alive and healthy. Three other children, one died of cholera infantum. Born at term ; instruments not used, labor easy. “ Nervous ” at birth. At about nine months began to cry out at nights and would draw up both feet as if in great pain. Never stood alone or walked. Now with assistance can walk a little. Cannot read ; mother says can sing. Cannot speak at all distinctly ; voice very thick. Sits bent over double, head hanging on chest, cannot sit upright for more than a moment. Moves head and body constantly. Knee-jerk marked. Ankles, knees, hips, stiff. Feet turned out at ankle ; knock-knee. In walking, puts toe to ground first. Pupils equal. Expression idiotic. Neck large, circumference fourteen inches. Trapezius and sternocleido muscles much enlarged. Tongue long. Teeth, two upper incisors a little chisel-shaped. Lower teeth well formed. Head diameters : biparietal, five and one-quarter inches ; occipito-frontal, six and one-half inches. Hands strong, well developed. Heels much drawn up. Can flex legs and feet. Ankle-clonus present. Muscles of legs not so well developed as arms. Spine flexible, no scoliosis.

CASE 2.—Vera M—, æt. 13. Mother living ; father killed. Five children living, four dead. Two born dead, one by craniotomy. This child born at eight

months; forceps not used, but labor long and hard. Nursed at breast; spasms at four months and again at six. Never could walk alone until fourth or fifth year. *Present state*: Left leg twenty-four and one-half inches long. Right leg twenty-five and one-half inches long. Knee-jerk ++. No ankle-clonus. Gait spastic; cannot walk without crutches. Patella drawn above condyles of femur by quadriceps extensor. Dentition tardy. Arms normal. Head: biparietal diameter five and one-quarter inches; occipito-frontal, seven inches; circumference twenty and three-eighths inches. Bright mentally, but queer.

CASE 3.—Addison D—, æt. 9. M. 5, 342. Deformity first noticed at fourteen months. Has never walked, sits doubled up with spine curved. Thighs look large, owing chiefly to large development of fat. Thigh can with difficulty be flexed on abdomen, pelvis rises on flexing thigh; knee very stiff, but can be gradually overcome. Legs much wasted; knee-jerk +. Toes are flexed and he cannot move them. Foot everted. Creeps about and walks with assistance on the outer side of his feet. Head well shaped: biparietal diameter six inches; occipito-frontal, seven inches; circumference twenty-one inches. Intelligence good, can read. Distinct nystagmus, convergent strabismus. Teeth well formed. Arms normal. Electrical examination: muscles respond actively to faradization.

CASE 4.—John P—, æt. 4. I. P. 4, 4. Parents healthy. Had convulsions at ten months; paralysis of legs, which are rigid. Now creeps on knees, equinovarus of both legs; reflexes +.

CASE 5.—George N—. æt. 5½. I. P. 4, 20. Natural labor, never had convulsions. Whooping-cough at four and a half years. Christmas, 1883, began to get restless and uneasy. Was treated for worms, and passed a few. Then became weak in the legs. The right leg first became weak, then in January, 1884, the left. The weakness steadily increased. Now the legs are rigid:

spastic contraction of muscles of feet; talipes equinus; walks on toes. Is bright. Reflexes increased.

CASE 6.—Robert G—, æt. 4 years and 9 months. M. 6, 48. Only child. Born with instruments; cried for eight hours steadily after birth. In twelve hours a large lump was formed in left occipital region. This was lanced, and a clot of blood removed. Intelligence good up to fifteenth month. At two years of age said to have had tubercular meningitis. Intelligence impaired ever since. Never had a convulsion. Has a depression at anterior fontanelle, which has a hard base. Says about a dozen words; not particularly fretful. Knee-jerk +. “Lead-pipe” leg; both legs nearly equal in stiffness; no wasting. Progresses by lifting his weight on his arms and throwing his body forward; cannot walk. Arms are strong. Nystagmus present. Phimosis. Teeth are in a state of decay and discoloration.

CASE 7.—Frank W—, æt. 6. M. 6, 100. A large baby, born with instruments, very long labor. Only child. Mother contracted syphilis six weeks before childbirth, and had a suppurating bubo. Began to walk at eighteen months, but never walked well, always stiffly. Teething a little tardy. Improved in walking till two years ago, when he had several spasms in rapid succession for twenty-four hours, which left him weak. The spasms came on in A.M. about 3 o’clock, did not gain consciousness until 5 P.M. Had much fever; was ill a week. Talked and walked more since the spasms. Has had slight spasms since. Last May had a series of very severe spasms. No stiffness in arms. Legs both spastic, toes turned in. Falls often. Has a very peculiar stiff gait, toes on ground; some incoördination in hands, particularly the left; picks up objects with difficulty; fingers very clumsy. Stiffness of legs most in extension. Feet extended. No nystagmus. Has headache at times. Knee-jerk + on both sides; no clonus. Slobbers a good deal. Does not talk well, but says many words. A slight internal strabismus. Biparietal diameter five and

a quarter inches ; occipito-frontal seven inches ; circumference nineteen and seven-eighths inches.

CASE 8.—Stella H—, æt. 6. I. P. B., 73. Never had fits ; sickly from birth ; no note as to labor. Had a fall at twenty-two months. *Status præsens* : Limbs well developed. At times limbs limber, but often rigid. Leg flexed, toes turned very much in. Can move legs, but cannot stand without assistance. After some effort rigidity is overcome. Arms normal. Electro-muscular contractility normal.

CASE 9.—Mamie McD—, æt. 3 years and 2 months. M. 7, 75. Mother died of phthisis. Born at seven months, no instruments used, labor natural. Always weak ; measles

FIG. 4.



Position of child in walking.

three months ago ; very severe spasms and crying spells, unconscious fifteen to thirty minutes. Can stand alone for a minute ; can walk when supported, cross-legged progression and walks on tiptoe. No special stiffness except when walking. Knee-jerk + +. No ankle-clonus. Ankles inverted ; movements of hip-joints normal. Hands not affected. Has night terrors. Intelligence good. No nystagmus. After five months' treatment much improved and could walk alone quite well.

CASE 10.—Violet M—, æt. 1 year and 4 months. I. P. 4, 46. First child in twenty-three years of married life. Two miscarriages. Labor natural and quick. Child weighed four pounds at three and a half weeks. Fed from bottle. Cholera infantum. No fever, no convulsions. The hands and arms are thin and never have developed well. Motion with them is very much impaired; difficulty in grasping or holding an object. Can walk with difficulty. Head brachycephalic. Intelligence poor. Knee-jerk+. —February 15th, 1888, is four years old. Has not been able to sit alone; cannot walk. Can say a few words, “papa” and “mamma;” understands everything; looks bright; has intelligent smile. Bad-tempered; cries a good deal; dribbles much. Head: biparietal diameter five inches; occipito-frontal five and three-quarter inches; circumference seventeen and three-quarter inches. No squint; no nystagmus. Teeth decayed. Had a convulsion last winter. The mother says that the child is like a *rag*. Arms flaccid; hands open; cannot pick up things; muscles ill-developed; a little stiffness of right arm. Legs extended, stiff, moves them about; feet extended, bent with difficulty. Gets very stiff at times, could almost be lifted by the legs. Plantar reflexes good. Sensation good.

CASE 11.—Joseph J—, æt. 20, colored. Sent to the Infirmary from a distance, and no history could be obtained except that his parents are healthy and that the present trouble had lasted from infancy. Patient is a well-grown lad, and has been accustomed to help at farm work. He is weak-minded, and cannot give a very satisfactory account of his past condition. He has an intelligent-looking face, although the forehead is low. Speech is imperfect, clips the words, and it is often hard to understand him. He has not much education, but can read a little. The arms are well developed; uses the hands and fingers well. The legs are muscular, of equal length. When recumbent he can flex them at the knee- and hip-joint, though they are somewhat stiff and there

is adductor spasm. He walks with a spastic gait, the legs stiff, the thighs adducted, and the toes kept close to the ground. The legs cross at each step, as shown at Fig. 6. The entire foot is placed on the ground. The arms are used to maintain the balance, and the body sways from side to side with each step. The knee-jerk is much increased, the ankle-clonus can readily be obtained ; superficial reflexes also increased.

The condition of the legs in these cases is practically identical with that in the spastic diplegias, and there are instances which link the two groups together. Thus, in Case 2 there was inability to use the hands properly, and the right arm at times became stiff, and in Case 10 the elbows were occasionally stiff, though at the examination there was no trace of it.

That in spastic paraplegia the cerebrum is less profoundly affected is shown by the greater number of children who are bright and intelligent and who ultimately learn to walk. Of the eleven cases three were mentally well developed ; the others presented various grades of feeble-mindedness. Six could speak plainly ; in four articulation was defective, and Case 10 could only utter a few words. Only one case had strabismus, and in two nystagmus was present.

In many cases the history was defective. In only three were there difficult labors, two requiring the forceps. The paramount influence of abnormal conditions of birth or of parturition in producing this condition is well illustrated in Little's cases, of which I think twenty-four can be selected as paraplegic. Of these in twenty-three there was either difficult labor or premature delivery ; six were first children. In only one instance were the forceps applied, but we must remember that in the fourth and fifth decades of this century the forceps were not used so often as they are at present. In none of Little's cases did the feet present, a point observed by Ross¹ and others.

The stiffness of the legs may not be noted for some

Loc. cit.

months after birth, but usually on washing and dressing the child the mother notices the rigidity. The child is late in attempting to walk, and then the awkwardness and stiffness of the legs become more evident. When standing the attitude is most characteristic—there is talipes equinus, varying from the slightest raising of the heel to a position in which the child actually stands on tiptoe. The heels are usually everted and the knees approximated, owing to the spasm of the adductors, which, with the gastrocnemii, become hard, tense, and prominent. In most cases, owing to the elevation of the heels, the body is supported on the balls of the toes. Fig. 5 represents the foot-marks in Case 9 as the child walked, with the soles chalked on a black surface. In other instances, as shown at Fig. 6, the entire foot may be set down at each step. Owing to the extension of the limb as it is being moved forward to take a step, the toes do not always clear the ground, but drag, so that the shoe-caps are usually much worn. The strong adductor action produces a remarkable crossing of the legs, and each foot is dragged over and planted in front of, or even to the other side of, its fellow. This is very well illustrated in the figures. When extreme, as in Fig. 6, which is taken from Case 11, the body is thrown from side to side in walking, and the arms are kept apart to help maintain the balance. In some instances the adductor spasm is so great that the thighs rub at each step, and one foot is shuffled before the other in a series of extremely short steps. The trousers may be rapidly worn out at the inner aspect of the knees in consequence of the constant friction. In attempting to flex the legs there is marked resistance, which gradually yields, and the limb can be bent, as Dr. Weir Mitchell has expressed it, like a bit of lead-pipe. The term “lead-pipe” contraction is often used at the clinics to designate this condition. The adductor spasm may be so extreme that it is impossible to separate the thighs—clasp-knife rigidity. When at rest the spasm may relax, but any attempt at movement or

FIG. 5.—Showing spastic gait in Case 9.

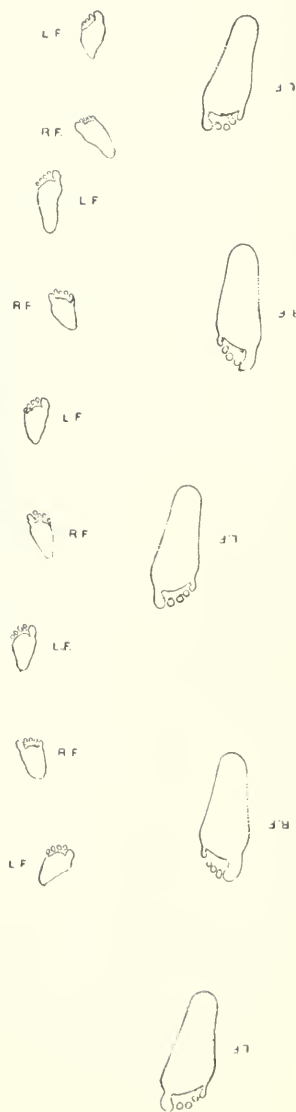


FIG. 6.—Illustrating cross-legged progression in Case 11.



an effort on the part of the doctor to flex the leg will at once induce it. Ultimately there may be constant flexion of the legs, with permanent contracture.

The reflexes are increased. The knee-jerk is almost invariably present, though in very young children it is sometimes not an easy matter to elicit. The ankle-clonus, as a rule, is not to be obtained. It was present in Cases 1 and 11.

Sensation is not impaired. Trophic lesions were not noticed. The functions of the bladder and rectum are unimpaired.

In brief, the symptoms of this affection in children are almost identical with those of adults, and the earlier writers on spastic paraplegia classed the cases together. Heine long ago expressed the opinion that these cases in children are of cerebral origin, and this is now generally accepted on the following grounds: 1. The frequent co-existence of symptoms indicating cerebral defects, such as idiocy, imbecility, nystagmus, and the like. 2. The occurrence of cases of bilateral spastic hemiplegia, in which the paraplegic symptoms are identical with those of tabes spasmodique—cases, moreover, in which the evidence is usually very clear of the existence of profound cerebral disturbance. All gradations are met with, from pure spastic paraplegia with perfect use of the arms, to instances of the most extreme bilateral spasm with or without disordered movements. 3. The paraplegic and diplegic cases present identical histories, and a large proportion of the cases in each group are birth palsies, the result of abnormal parturition. 4. As the diplegic cases have been shown to depend upon symmetrical disease of the motor areas with, in many instances, descending degeneration, the conclusion has not unnaturally been drawn that the paraplegic form was the result of a similar though less extensive lesion. Ross, Hadden, and Gowers take this view; d'Heilly thinks that there is not necessarily involvement of the brain, but that it may be a primary degeneration of the lateral tracts.

THE MORBID ANATOMY of infantile spastic paraplegia remains to be worked out. In a tolerably careful review of the literature, I can find but one record of a carefully performed section in a case of this kind. Förster,¹ in a report from the Dresden Children's Hospital, among the spastic palsies, gives the following case: Boy, æt. 2. No history of the parents or of the delivery. At the age of one and a half years the paralysis was noticed, and he began to use the few words of speech which he had acquired. When seen at about the age of two, the arms were normal, the legs stiff and with strong adductor spasm. When attempting to walk he stood on the outer aspects of the feet with the heels raised. Knee-reflexes exaggerated. Intellect feeble. The case appears to have been one of true spastic paraplegia. The post-mortem, by Birch-Hirschfeld, showed a moderate grade of general cortical sclerosis, with slight dilatation of the ventricles. The basal ganglia were normal; no note as to pyramidal tracts in crus or medulla. The cord was firmer than normal, and the lateral tracts presented a gray-white, translucent aspect, which was regarded as a descending degeneration, the consequence of the brain lesion.

DIAGNOSIS.—The diagnosis of spastic diplegia and paraplegia is usually easy, but there are cases from which they must be carefully distinguished and which, at times, closely simulate them. The condition described by writers on children's disease as *idiopathic contraction with rigidity*, *tonic contraction of the extremities*, the *contractures* of the French writers, is very apt to be confounded with true spastic paralysis and *vice versa*. The confusion which exists is illustrated in several recent papers, notably those of Onimus² and Launois³ and the thesis of Simard⁴. The majority of the cases reported

¹ 'Jahrbuch für Kinderheilkunde,' Bd. xv.

² 'Revue men. des maladies de l'enfance,' 1883.

³ 'France Médicale,' 1884.

⁴ 'Thèse de Paris,' 1884, No. 85.

by these writers belong to the category of spastic paralysis due to cerebral disease.

The chief difference between these conditions may be thus tabulated :

<i>Pseudo-paralytic rigidity.</i>	<i>Spastic paralysis; di- and paraplegia.</i>
Follows a prolonged illness. Is often associated with rickets, laryngismus stridulus and the so-called hydrocephaloid state.	Usually exists from birth. History of difficult labor, of asphyxia neonatorum or of convulsions.
Begins in hands as carpo-pedal spasm; often confined to hands and arms.	Arms rarely involved without legs, and not in such a marked degree.
Spasms painful and attempts at extension cause pain.	Usually painless.
Intermittent and of transient duration.	Variable in intensity but continuous.

The history, the limitation in many instances to the arms, the existence of rickets or other constitutional disturbance, render clear the diagnosis. The spasm in idiopathic contraction may be extreme, the arms adducted, the forearms strongly flexed and the hands clenched on the chest. In none of the cases of spastic diplegia have I met with such inflexible rigidity of the arms as existed in a rickety child which I saw with Dr. Major, of Montreal. From tetany, which in children is closely related to the carpo-pedal spasm, and occurs under similar conditions, the distinction would rest largely on etiological considerations. In early life the association with rickets and diarrhœa, and the greater involvement of the upper extremities, are features of tetany.

It must not be forgotten that tumors of the pons and of the cerebellum may produce a bilateral rigidity when the motor paths are involved or compressed. Tubercular growths of the cerebellum seem specially liable to induce this symptom. In the Gulstonian Lectures for 1886, Sharkey¹ reports four cases, in each of which the tumor occupied such a position that it compressed either pons or medulla.

¹ 'Spasm in Chronic Nerve Disease,' London, 1886.

CHAPTER V.

PATHOLOGY AND TREATMENT.

PATHOLOGY.—Varied as is the anatomical condition, the lesions have this in common—interference with the motor centres, or with the conducting paths of the cerebrum. In reviewing a large number of post-mortem records, or in studying such a series of brains as that prepared by Dr. Wilmarth, at Elwyn, we are impressed, on the one hand, with the extent to which sclerotic and other changes may exist without symptoms if the motor areas are spared, and, on the other hand, with the degree of permanent disability which may exist with even slight affection of this region. Our knowledge is so limited to the appearances and states years after the onset of the symptoms, the final results of processes long past, that we are scarcely in a position to discuss accurately, in all its aspects, the pathology of this interesting group of cases. It is something, however, to get an outline for our ignorance and to ascertain in which direction facts are needed to sustain, or, it may be, to upset our theories.

A certain number of the cases of hemiplegia in children are due to *hæmorrhage* from causes identical with those which prevail in the adult, and I have given illustrative instances of true apoplexy with laceration of the cerebral substance; but it may be safely concluded, I think, that hæmorrhage is not the common cause, and accounts for a very small percentage. A small proportion of the cases of hemiplegia come under the designation of birth palsies, as there is a history of persistence of the paralysis

from birth, and of the occurrence of difficult labor, often necessitating the application of forceps.

In the cases of *birth palsy*, which result usually in bilateral hemiplegia or paraplegia, the evidence points strongly to meningeal hæmorrhage as one of the chief causes of the disorder. The great majority of these, as we have seen, present, at birth, one of two conditions, asphyxia or convulsions. The children are resuscitated only after prolonged attempts at artificial respiration; more commonly convulsions occur, either immediately after birth, or within the first ten days of life. Facts have been gradually accumulated to show that hæmorrhage, usually meningeal, is a very frequent condition in children dying shortly after birth of asphyxia, or convulsions, and as the birth palsies almost invariably have this history it seems reasonable to conclude that, in the cases which recover and subsequently present signs of motor disturbance, a similar, though less intense, lesion has existed.

Apoplexia neonatorum is by no means an uncommon event. Little fully understood its importance, and quotes from Cruveilhier, Evory Kennedy, Hecker, and Weber to show the occurrence of meningeal hæmorrhage and capillary extravasations in newborn children. In 1880, Litzmann¹ communicated the results of the examination of 161 newborn children, in eighty-one of whom the spinal canal was exposed. There were thirty-five instances of meningeal hæmorrhage, in nineteen the extravasation being in considerable amount. The exudation existed in both cerebral and spinal meninges.

Parrot,² in thirty-four cases of cerebral hæmorrhage in the newborn, found five in which the blood was in the cavity of the arachnoid, and twenty-six in the subarachnoid space. In a large proportion of the cases the extravasation was bilateral. This author makes no allusion to the association of the lesion with abnormal labor.

¹ 'Archiv f. Gynäkologie,' Bd. xvi.

² 'Clinique des Nouveau-nés,' Paris, 1877.

Dr. Sarah J. McNutt has reported ten cases,¹ and her paper is very convincing as to the frequent association of this condition with abnormal labor and with asphyxia and convulsions in the newborn. It may occur when the parturition has been normal, and may be deferred some days, or even weeks, as in the following case, which occurred in Dr. Parvin's wards at the Philadelphia Hospital, and the specimens from which I saw with Dr. Stahl.

Child, æt. 6 weeks, mother single; labor normal. Remained healthy until the forty-fifth day, when at 8 A.M., it had a convulsion, at first on the right side, and finally general. Convulsions were repeated through the day and an ecchymotic rash appeared on trunk and extremities. Death at 10 P.M. The post-mortem showed extensive meningeal hæmorrhage, a large clot over the cortex, particularly on the left side. There was much more blood in the left than in the right Sylvian fissure, and it extended under the frontal convolutions; it looked as if the bleeding had begun here, but the most careful examination of the vessels showed no changes in the arteries or thrombi in the veins. There were clots also in the posterior and middle fossæ, and a uniform sheeting of coagulated blood extended beneath the spinal dura.

I saw a second case, this winter, at the Philadelphia Hospital, and I am indebted to Dr. Hirst, who made the autopsy in both these cases, for the following notes: Case of twins; first child delivered with forceps—head presentation. The second child presented the breech, and there was great difficulty in the extraction of the head, required much force. Twenty minutes after birth, left lateral convulsions occurred and were repeated at intervals. The child lived forty-eight hours, having become intensely anæmic. The post-mortem showed the viscera normal. The brain, in places, was remarkably soft, the tissue almost diffuent. A large clot existed beneath the dura mater on the right side, very thick over

¹ 'American Journal of Obstetrics,' 1885.

the cortex. It dipped into the fissures and sulci and extended down the cord. About the middle of the longitudinal sinus there was an irregular laceration, several lines in length.

The extravasation has in many cases been thickest over the motor areas, and from what we know of the changes which time may effect in effused clot, there is nothing inconsistent in the belief that sclerosis from compression or porencephalus from destruction might ultimately result. Gowers advocates this view in the case of the birth palsies, which, indeed, he considers in his work under the heading of "Infantile Meningeal Hæmorrhage." Probably all the cases cannot be assigned to this cause, and I think, with Jacobi,¹ that certain of them may be due to foetal meningo-encephalitis. Certainly in Ashby's case,² in which, in a child twenty-two months old, the cortex was smooth, without a trace of convolutions, the sclerosis must have begun during embryonic life.

The frequency of spinal hæmorrhage renders it not improbable that some of the cases of spastic paraplegia may be due to this cause; and Ross suggests that as severe traction in feet presentation has been known to tear the cord, slighter degrees might injure the pyramidal tracts, and lead to a sclerosis.

We have then in the spastic diplegia, and in a few cases of hemiplegia—the true birth palsies—information which enables us to assign to hæmorrhage an important rôle. More positive knowledge may ultimately be obtained by the dissection of cases at different ages after the onset.

When we turn to the cases of *infantile hemiplegia*, which come on during the first two or three years of life, we are met at once with conflicting theories. It is well to bear in mind that we are called upon to explain the

¹ Discussion at the Academy of Medicine, New York, January 25th, 1888. 'N. Y. Med. Record,' 1888, i.

² Loc. cit.

mode of origin of sclerosis and porencephalus, the two conditions present in the great majority of the cases. A certain number of cases of infantile hemiplegia are due to hæmorrhage, to embolism and aneurism, a few to tumour, as glioma or chronic tubercle; but, as we have seen, these form a fractional part. We require to know the pathological process lying at the basis of the convulsive attacks with coma, which come on suddenly, or after a slight febrile movement, frequently succeed an infectious disease, and leave a hemiplegia with too often its disastrous sequences—epilepsy and imbecility. In a large proportion of the cases the disease is such a clinical unit, with symptoms as marked and definite as those of infantile spinal paralysis, that we might expect a corresponding uniformity in the anatomical lesion. Unfortunately we are, so far as I can ascertain, entirely without information upon the state of the brains of children dying during or shortly after the attack; and the question resolves itself into an explanation of the conditions most commonly met with years after the onset, viz. sclerosis and porencephalus.

A few years ago Strümpell¹ suggested that the lesion was the cerebral counterpart of the infantile spinal palsy, a *poliencephalitis* of the motor areas of the cortex, analogous to the poliomyelitis of the anterior horns. The two affections are very similar in the mode of onset, in the age affected, and in the liability to follow one of the infectious diseases. The more frequent occurrence of convulsions and the more prolonged coma might be expected from the seat of the disease. This very plausible and suggestive view has not met with much favor, not so much, I think, as it deserves. Against it has been urged the absence of anatomical facts, but this defect it shares with several theories. I see no improbability in the view; and it is possible that we shall have, ere long, the necessary anatomical facts to support it. The forms of encephalitis which we know: Virchow's encephalitis of

¹ Loc. cit.

the newborn; the miliary encephalitis due to septic causes, such as has been found in diphtheria and in aphthous stomatitis; and the steatosis of Parrot, arise under different circumstances, and do not present the symptoms of these cases of infantile hemiplegia. J. Lewis Smith has suggested¹ that certain of these cases of infantile hemiplegia are due to cortical encephalitis, set up by the poison of cerebro-spinal fever.

The frequency with which the disease is associated with or follows one of the infectious diseases is suggestive. Endocarditis is not rare in scarlet fever, measles and diphtheria, the affections which most commonly precede the hemiplegia, and embolism may account for a certain number of these cases. In diphtheria there may be plugging of the smaller cerebral arteries with micrococci, without the occurrence of endocarditis. There is another way in which the relation of the infectious diseases may be regarded. We know that in certain fevers, typhoid particularly, changes have been met with in the smaller arteries leading to anæmic necrosis in the corresponding districts. This has been worked out in the heart by Landouzy and Siredey,² and if it occurs in limited areas of this organ it might also take place in the brain. From what we know of sclerosis in other organs, notably the heart, the rôle played by the blood-vessels in the process is all-important. Kundrat has already suggested that porencephalus results from an anæmia of definite areas corresponding to arterial distribution, but without arterial lesion. It is difficult to conceive of such a condition, but a widespread endarteritis, similar to, but more extensive than, that which is known to induce anæmic necrosis of the heart-muscle might initiate a sclerosis; or, if the obstruction was suddenly effected, and large vessels involved, produce a rapid necrosis, the final changes of which would represent porencephalus. That this latter condition results from vascular obstruction we

¹ 'Medical Record,' 1887.

² 'Revue de Médecine,' 1885.

have evidence in the large defects which are occasionally found in brains of adults, and in the one organ there may be regions of softening in all stages of regressive change. Less abrupt processes in smaller vessels may account for certain of the cases of sclerosis, and the changes which Martin, Sabourin, and other French writers have found in the smaller arteries in cirrhotic regions, make it by no means improbable that the cerebral counterpart has also a vascular origin.

Jendrassik and Marie¹ have described vascular lesions in cerebral sclerosis, which they believe to be primary. The change is in the perivascular region, which is greatly increased in size and intersected by connective-tissue growth, which is in intimate relation with the vessel wall and with the neuroglia. While regarding the periarteritis as the primary change in the cerebral paralysis of children, these authors do not exclude embolic and thrombotic processes in the smaller vessels. They refer also to the connection of these with infectious processes.

In a third way, too, an infectious disease might induce hemiplegia, by causing changes in the cerebral motor centre similar to those which occur in the disseminated myelitis following measles and scarlatina. We have not had until recently much positive knowledge of the latter, but such a condition as Barlow² has described in the cord shows the direct influence of the specific poisons on the elements of the gray matter. May not this occur in the motor cells of the cortex as well as in those of the cord? But here we return to Strümpell's view. The changes in the gray matter are, in most of these instances, necrotic, rather than inflammatory, and the vascular lesion is the primary one. In his researches in sclerosis Adamkiewicz³ found the destruction of the nerve elements the first step in the process, to which the proliferation of the neuroglia was secondary.

¹ 'Archiv de Physiologie,' 1883.

² 'Medico-Chirurgical Transactions,' vol. lxx, 1887.

³ 'Neurologisches Centralblatt,' 1886.

In favor of the embolic theory it may be urged that the affected areas correspond with definite territories. Of porencephalus this is to a great extent true, but not always of sclerosis, which may be most irregularly distributed. I purposely noted, when discussing the morbid anatomy of sclerosis, the districts involved in the ten specimens in the Museum of the Elwyn Institution. The involvement of two or three regions, far apart from each other, and the frequency with which the territory of the posterior cerebral artery is the seat of the sclerosis, are against embolism as a cause.

Gowers has suggested that the lesion in these cases is *thrombosis of the cerebral veins*, a well-recognized condition in children, which sets in with convulsions. Usually there is also sinus thrombosis. Certainly, there are no veins in the body so favorably situated for the coagulation of blood; "they are roomy in proportion to the amount of blood they carry, they are tortuous and abundantly anastomosing, so that the current of blood is almost reversed at some points and can easily stagnate" (Edes).¹ Gowers quotes a case of Money's in which the thrombosis occurred after scarlet fever. Handford² has reported the case of a boy æt. 5, in whom the cerebral veins were like whip-cords, and there was hæmorrhage on the surface of both hemispheres. There was also sinus thrombosis. The symptoms, however, were not those of infantile hemiplegia. Parrot³ states that this condition is very common, and in plate iv of his work there is a beautiful illustration of thrombosis of the surface veins with hæmorrhage. The advantage of this view is that we are dealing with recognized lesions, of common occurrence in infancy, capable of explaining satisfactorily the symptoms; and yet the proof is lacking, the demonstra-

¹ 'System of Medicine by American Authors,' vol. v, p. 982, Philadelphia, 1886.

² 'British Medical Journal,' 1887, i, p. 1098.

³ Loc. cit.

sion of its existence in one of these cases of infantile hemiplegia has still to be made.

To sum up: Infantile hemiplegia is probably the result of a variety of different processes, of which the most important are:

(1) Hæmorrhage, occurring during violent convulsions or during a paroxysm of whooping-cough.¹

(2) Post-febrile processes: (a) embolic; (b) endo- and peri-arterial changes; and (c) encephalitis.

(3) Thrombosis of the cerebral veins.

There are several problems of interest in connection with the pathology of infantile spastic paraplegia. Of special importance is the condition of the pyramidal tracts in the cord, and the attention of future observers should be directed to this point. In three cases of spastic diplegia, and in the one case of spastic paraplegia, there was descending degeneration. In Ross's case the cord was normal. In Ashby's case was the change in the pyramidal tract truly a degeneration, or was it not rather arrest of development? Most probably the latter, in connection with a brain the cortex of which was smooth and presented no trace of convolutions. In Ross's case, though the cord was said to be normal, yet the anterior pyramids were not more than half the normal size, and the lateral columns were smaller. Alexandra Steinlechner² has reported the case of a lad named Post, æt. 6, helpless from birth. The legs were in equino-varus position, but as there was no note of the condition of the arms, I did not know in which category to place the case; it was probably one of spastic diplegia. Post mortem, bilateral porencephalus was found; the lateral tracts were undeveloped and did not contain more than one half the normal number of nerve-fibres. Hervouet³ has recorded

¹ Dr. Samuel West reported a case of this kind to the Clinical Society of London last year ('Medical Press and Circular,' 1887).

² 'Archiv für Psychiatric,' Bd. xvii.

³ 'Archives de Physiologie,' 3e série, tome iv, 1884.

the case of an idiotic child, æt. $3\frac{1}{2}$, without spasms or contractures, in which there was a condition of microcephalus with complete absence of development of the pyramidal tracts in the cord. The motor strands in the cord do not attain their full development until after birth, according to Hervouet not until the third or fourth year. It is a question of no little interest to determine the exact condition of these tracts in cases of diplegia and paraplegia of children. Sharkey¹ states that in cases of congenital absence of the whole or part of the motor centres of the brain, which are accompanied by contracture, the pyramidal tract has proved to be deficient. And yet in Hervouet's cases these strands were undeveloped without any spasm of the muscles. It would really appear then that actual sclerosis of the lateral tracts is not an essential condition of spastic rigidity though present in a great majority of the cases. In the adult, in the cases of Schultz,² Strümpell³ and Anna Klumpke⁴, the pyramidal tracts have been found normal in spastic paralysis, and Blocq⁵ in his recent monograph acknowledges that contractures may exist without affection of these parts. The problem of rigidity is by no means an easy one to solve. The over-action of the spinal centres seems the most reasonable view, at one time conditioned by the suppression of the functions of the pyramidal tracts, the absence of inhibition, the "let go" theory of Hughlings Jackson, at another possibly excited reflexly, even when the motor strands in cords are intact.

We shall need in future autopsies on cases of spastic paraplegia in children specific details as to the condition of the lateral tracts, and particularly as to degeneration or absence of development. The two conditions might

¹ Loc. cit., p. 11.

² 'Deutsches Archiv f. klin. Med.,' Bd. xxii.

³ 'Archiv f. Psychiatrie,' Bd. x.

⁴ 'Dictionnaire Encyclopédique,' art. "Tabes Spasmodique," Raymond, 1885.

⁵ 'Les Contractures,' Paris, 1888.

readily be confounded as can be gathered from an examination of the plate which illustrates Hervouet's paper.

TREATMENT.—In the majority of the cases the physician is called at the onset to treat an attack of convulsions or of coma, developing suddenly or after a few days' indisposition, or following, perhaps, one of the fevers. The paralysis is not apparent until the active symptoms have subsided, though if the convulsions are severe and unilateral it may be anticipated. These symptoms demand the bromides with chloral, a calomel purge, cold to the head, and, if necessary, leeches. The cases resemble so closely the ordinary convulsions of infancy associated with teething that the development of so serious a condition as hemiplegia is a great shock to the parents, who are very apt to blame the physician for having done too much or too little. In such an accident following vaccination the doctor is fortunate indeed if he escape unjust criticism. In the birth palsies, unless there are marks of the forceps, the condition of the limbs of the child does not attract attention, and it may be weeks or months before the disability is discovered.

The traumatic cases, such as the three reported, suggest surgical interference, two of them at any rate, as they were doubtless due to hæmorrhage; but it is well to remember, in these days of safe surgery, that in children traumatic hemiplegia may sometimes disappear completely in a few days. I reported an instance some years ago¹ of a child, æt. 23 months, who had fallen from a balcony, and was admitted to my wards with a large hæmatoma of the scalp, left hemiplegia and coma. The symptoms gradually disappeared and the child made a complete recovery. A case was narrated to me by Dr. Murray Cheston of a child who was tripped by his brother and fell on his head. There was no external wound and no fracture. Hemiplegia developed, which in a few days disappeared completely, and the boy is now quite well.

¹ 'Canada Medical and Surgical Journal,' vol. viii.

When the paralysis is established medicines are rarely called for, and the indications are to favor the natural tendency of the hemiplegia to improve and to lessen the rigidity and contractures, and, if necessary, overcome them by mechanical or surgical measures.

While it is impossible to predict, in any given case, to what extent the original palsy will disappear, we can usually expect a return of power in the leg and face, incomplete in the former, often perfect in the latter. In rare instances, two only in the series of one hundred and twenty cases, did the paralysis entirely disappear. The residual palsy is most marked in the upper extremity, and even when the arm is tolerably useful the fingers do not regain the power of delicate movements. Temporary arrest of growth and some degree of atrophy follow invariably, but as the child grows up we rarely see the great discrepancy in size of members which is common after the spinal infantile palsy. To maintain the nutrition of the paralyzed parts we employ warmth, massage, and electricity. The temperature of the affected members is usually lowered and the circulation sluggish, so that the limbs should at first be wrapped in cotton-wool, and when the patient begins to make efforts at walking, flannel underclothing should be used. Attention to this apparently minor point should be impressed upon the mothers; it is too often neglected. Massage of the paralyzed muscles should be practised daily. Simple directions should be given to the mother or nurse, in the absence of a professional rubber. The chief point is to manipulate the muscles thoroughly, and I usually order the rubbing to be done with sweet oil, morning and evening, for twenty or thirty minutes. I have strong belief in the efficacy of this treatment in keeping up the nutrition of the enfeebled muscles. To be of any service the applications must be continued for months.

Electricity is probably of no service in the original lesion, which may be left to nature, undisturbed by galvanism, which has been recommended by some authors.

Of positive benefit is faradization of the paralyzed muscles, which must, however, be carried out persistently for months. Next to the rubbing, it is the most important agent upon which we rely to prevent atrophy and maintain the nutrition of the parts. It is not often that we find hospital patients able to spare the time for the prolonged treatment needed in these cases. Fortunately, the wasting is not extreme, and even without the treatment the leg and face recover more or less completely.

A considerable number of the cases come under observation when there is marked rigidity and contractures. For this condition in the palsied arm of hemiplegia not much can be done, but in the cases of spastic diplegia and paraplegia manipulations and surgical measures will often enable a crippled patient to walk. Persistent massage with strong flexion and extension of the limbs, as recommended by Dr. Weir Mitchell is of great service. Case 9 of the spastic paraplegia cases could not walk when brought to the Infirmary, and after about a year's persistent treatment—frictions with oil and forcible flexion and extension—she can now get about the house quite well. Sooner or later the majority of these cases come under the care of the orthopædic surgeons who were dealing with this condition—and some indeed, as Heine, knew upon what it really depended—several decades before physicians had recognized that it was primarily an affection of the nervous system. The woodcuts and plates given in the works of Heine, Little, and Adams show how well they dealt with these cases, and we have many illustrations at the clinics of my colleagues, Drs. Morton, Hunt, and Goodman, of the good results of division of tendons and of the application of proper apparatus.

There are one or two special symptoms calling for comment. The aphasia, as we have seen, usually disappears, but it may be helped by systematic education, and these cases do better and recover more promptly than aphasia in the adult.

Epilepsy is a distressing symptom for which many of the cases seek relief. It is well to recognize clearly the cortical nature of the attacks, and let the parents know that a cure can rarely be anticipated. It is encouraging, however, to note that the seizures may lessen greatly, and prolonged periods of quiescence are not uncommon. The attacks of hemi-epilepsy without loss of consciousness may persist for years before a general convulsion with loss of consciousness occurs. In some instances the attacks are repeated with extraordinary frequency, twenty or more daily. In the transient attacks of *petit mal* the bromides do good; but, as a rule, in the true Jacksonian fits, unless there is much irritability and excitement, I have not found them very beneficial.

An important question of practical moment has arisen in connection with the propriety of surgical interference in these cases. Jacksonian epilepsy has now its surgical aspects, and there have already been several successful cases of removal of tumors from the motor areas of the cortex. Infantile hemiplegia offers some of the most typical instances of cortical epilepsy, and it may be well to consider how far it is likely that surgical interference can here be successful. As we have seen, in the review of the morbid anatomy, the conditions to be dealt with are (1) apoplectic, embolic or thrombotic foci, which are as frequently in the territory of the central as of the cortical arteries; (2) sclerosis; and (3) porencephalus. I do not think that, in any of the cases which I have reviewed, the anatomical condition offered the slightest possibility of relief from surgical interference. I except the case of glioma of the paracentral lobule to which I referred. This girl had had Jacksonian epilepsy for many years and the tumor could have been readily removed. In the second autopsy of the Elwyn cases, in which there were blocks of sclerosis, the mass causing the hemiplegia might have been removed, but there were several other areas.

There have been two cases of operation in the cortical epilepsy of infantile hemiplegia. Bradford, of Boston,

operated on a case of Bullard's¹. A boy, æt. 4, forceps delivery, idiotic, and in a state of right spastic hemiplegia. There was a scar and depression on the right parietal bone, due probably to the forceps. The trephine was first applied over this part, but the brain-substance beneath was found normal. The left side was then trephined and a porencephalous condition of the Rolandic region found. The child died the next day. No autopsy was allowed. The other case was operated on in the Infirmary by Dr. Morton. I have referred to it when speaking of the symptom of epilepsy (Case 97). Dr. Mitchell, under whose care the child was, believed there was a possibility that the lesion might be a localized area in the motor zone, which could be removed, and in this opinion Dr. Sinkler and I concurred. Accordingly Dr. Morton exposed the Rolandic region and found an œdematous condition of the membranes, but no focal disease. Whether the portion removed is sclerotic has not yet been determined. The child recovered perfectly from the operation, the dressings were removed on the sixth day. The spasm of the arm has lessened and the attacks of epilepsy have reduced in frequency.

There are several circumstances which militate against the probable success of operations of this kind. The nature of the lesion is such that not much can be anticipated. When sclerosis exists the area is usually too large for removal; when porencephalus is present, we are not, as Dr. Morton Prince said in the discussion on Dr. Bullard's case, "likely to improve a pure paralysis due to a hole in the brain, by making the hole bigger." But a more serious objection was raised in the same discussion by Dr. J. J. Putnam, viz. the existence in these long-standing cases of descending degeneration. The paralysis in such instances could not be benefited, and it is only in exceptional instances that we could expect the epilepsy to be relieved. Time alone will determine how far in the removal of centres for cortical epilepsy the im-

¹ 'Boston Medical and Surgical Journal,' 1888, vol. i.

provement is temporary or permanent, and how much less irritating the scar of a surgical wound will be than the cicatrix of an accidental trauma.

More serious in many respects, and more distressing to friends and relations, are the mental defects so apt to be associated with these cerebral palsies. The subjects of bilateral hemiplegia are usually imbecile, often idiotic; the spastic paraplegic cases offer greater hope of mental improvement, and many of the patients are intelligent and learn to read and write. They rarely suffer from epilepsy, so potent a factor in inducing mental deterioration. A large percentage of the cases of hemiplegia grow up feeble-minded, a larger percentage than indicated in our list. Much depends, no doubt, upon the area and regions of cortex involved, but even in such localized lesions as that shown at Fig. 1, there may be serious mental deficiency, showing that the actual damage is more than is apparent. The injury to the brain is usually done at the very time when the faculties are developing and the education of the senses is preparing the way for the higher intellectual processes. It is not surprising that in so many cases the damage is irreparable and that idiocy or imbecility results. A few years ago these cases were neglected and thought incapable of education. The results which are attained by Dr. Kerlin and his staff, at the Pennsylvania Institutions for Feeble-minded Children, and in other public and private establishments, demonstrate that the lesson which Dr. Seguin, Sr., and others labored so hard to teach has been well learned—the lesson that with patient training and kind care many of these poor victims may be rescued from a condition of hopeless imbecility and reach a fair measure of intelligence and self-reliance.

4114

